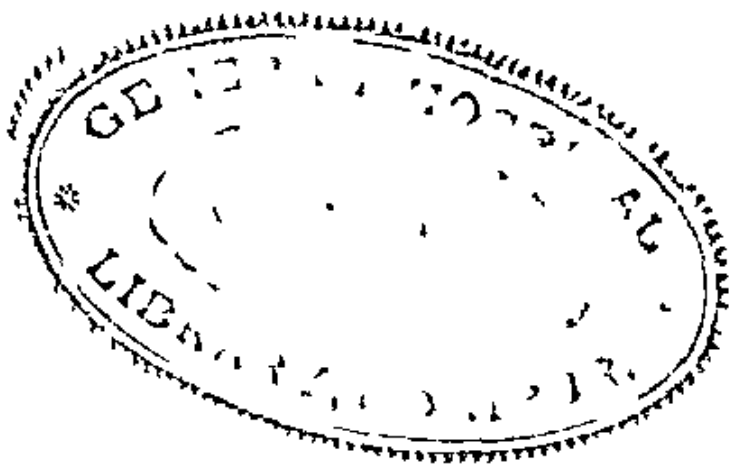

SURGICAL TREATMENT FOR ABNORMALITIES OF THE HEART AND GREAT VESSELS

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Edited by

Michael E DeBakey, M D

Associate Professor of Surgery
Tulane University
New Orleans, Louisiana

and

R Glen Spurling M D

Clinical Professor of Surgery
University of Louisville
Louisville, Kentucky

THORACIC SURGERY DIVISION

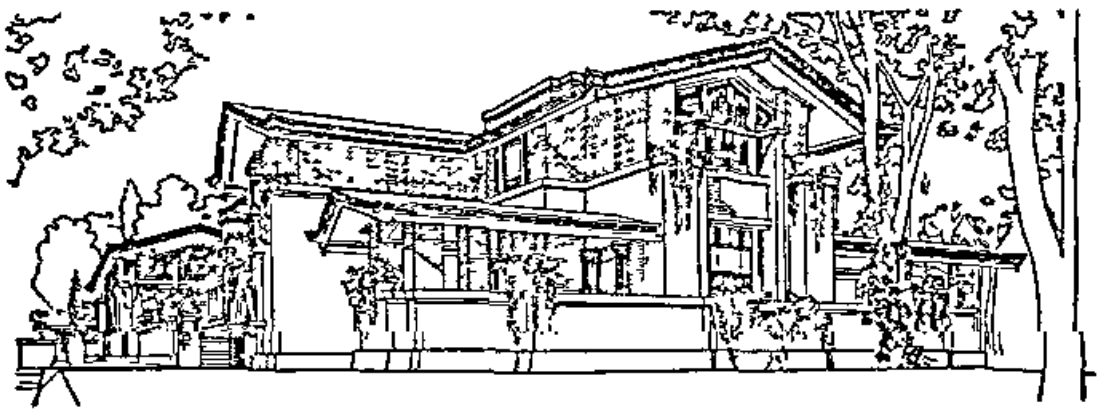
Editor Brian Blades, M.D

Professor of Surgery
George Washington University
Washington D C

SURGICAL TREATMENT FOR ABNORMALITIES OF THE HEART AND GREAT VESSELS

By ROBERT E. GROSS, M.D.
William E Ladd Professor of Child Surgery
Harvard University Medical School
Boston, Massachusetts

THE BEAUMONT LECTURE
WAYNE COUNTY MEDICAL SOCIETY
DETROIT, MICHIGAN



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PREFACE

Much of the subject material which is gathered here for the Beaumont Lecture of 1946 has been presented elsewhere, or has been recorded in previous medical writings It is therefore necessary to reproduce illustrations from various journals, and this opportunity is taken for expression of thanks to the following periodicals for the use here of certain charts drawings, and photographs I am indebted to *The Journal of the American Medical Association* for Figure 6 from "Experiences with Surgical Treatment in Ten Cases of Patent Ductus Arteriosus," 115 1257, 1940, *The Journal of Pediatrics* for Figure 10 from "Surgical Closure of the Patent Ductus Arteriosus," 17 716, 1940, *The New England Journal of Medicine* for Figures 27, 28, and 29 from "Coarctation of the Aorta, Experimental Studies Regarding Its Surgical Correction," 233 287, 1945, and *Surgery*, for Figure 30 from "Surgical Correction for Coarctation of the Aorta," 18 673, 1945

ROBERT E GROSS, M D

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SURGICAL TREATMENT FOR ABNORMALITIES OF THE HEART AND GREAT VESSELS

INTRODUCTION

IT IS A PLEASURE to come before the Wayne County Medical Society* to deliver the annual lecture of the Beaumont Foundation I am fully cognizant of the honor which you have bestowed upon me, and I would like to express my appreciation to the Foundation Committee and to the medical society as a whole The list of previous holders of this lectureship includes physicians and scientists of the highest quality, and it is with great humbleness that I find myself grouped with them The subjects which have been presented to you in former years include a wide variety of topics in the basic sciences and in the clinical branches of medicine It is with some hesitation that I herewith call to your attention a rather small group of patients who have congenital malformations of the heart, the aortic arch, and the great vessels in the superior mediastinum Until a few years ago there was no definitive treatment for such individuals, but now therapeutic procedures are available for correcting some of these abnormalities. This field of surgical endeavor is in its infancy and would appear to be limited, but it is rapidly growing and has excited rather widespread interest Hence I feel justified in summarizing our recently acquired experiences because they not only relate what has been accomplished in the past but possibly they will stimulate others to devise methods for treating additional types of malformations of the cardio-vascular system

The field of medicine and surgery is becoming so complex that it is rare for therapeutic advances to come from the labors of one man It becomes increasingly apparent that we must depend upon the suggestions, the help the criticism and the many other contributions which are made by our confreres The work which is here summarized is no exception to this generality Through the years I have leaned heavily upon the guidance and support of my superiors and peers My interest in this field was developed by the constant prodding of Dr John Hubbard, who understood many of the problems of congenital heart disease and who was anxious

* The 25th Beaumont Lecture of the Wayne County (Mich.) Medical Society

to find a way for their surgical correction My deepest thanks are offered to the various chiefs who have given me a free rein in the conduction of laboratory experiments and in the treatment of human patients I am deeply indebted to Dr S Burt Wolbach the Shattuck professor of pathology to Dr Elliott B Cutler the Moseley professor of surgery to the late Dr Kenneth B Blackfan the Rotch professor of pediatrics and to Dr William E Ladd the former Ladd professor of children s surgery—all of the Harvard Medical School The roentgenologic studies on our patients were begun by Dr George Wyatt and were continued by Professor Merrill C Sosman and more recently by Dr Edward B D Neuhauser This surgical progress has had a broad foundation of experimental work in the Laboratory for Surgical Research at the Harvard Medical School and I owe a great debt to Dr Elliott C Cutler for the generous use of his laboratory facilities and for the encouragement which he has given from time to time Individuals with cardio-vascular anomalies often present variations in function and rich contributions have come from Dr C Sidney Burwell and Dr Eugene C Eppinger as a result of their studies on these disturbed physiological mechanisms Finally and of extreme importance has been the constant loyalty and enthusiasm of various members of the house staff of the Children s Hospital and the Peter Bent Brigham Hospital who have devoted a great deal of time and energy to the care of patients before and after operation and have thus made possible the results which are here recorded

The field of cardio vascular defects includes a wide variety of malformations There are the common and well known deformities but in addition there are many bizarre and rare types which have excited little more than academic interest It is obvious that a large number of malformations are beyond the possibility of surgical relief but this pessimistic angle should not alter the brighter side of the picture regarding those which can now be treated with a considerable degree of success It is on this latter group that I would like to focus your attention and to consider briefly what we have learned about each of these problems Of foremost interest to me has been the work on the patent ductus arteriosus and its surgical cure hence this will be considered in greatest detail Abnormalities of the cardiac envelope rarely give rise to difficulties yet I would like to present an individual with symptoms from a pericardial defect which were relieved by surgical means Within the last year there has come to light the exceedingly promising

work of Blalock and Taussig which brings relief to those who suffer from a tetralogy of Fallot It has long been known that anomalous arteries in the superior mediastinum can press upon the trachea or esophagus and interfere with their function, but now methods have become available for correcting some of the malformations Finally, obstruction in the main aortic pathway, so called coarctation of the aorta, appears to be amenable to surgical relief in some instances, and I would like to summarize our studies on this condition and its therapy

PATENT DUCTUS ARTERIOSUS

In fetal life the atelectatic state of the lungs and the consequent small size of the pulmonary vascular bed make it necessary for nature to provide some method whereby a large part of the blood can be kept circulating without passing through the lungs This function is fulfilled by the ductus arteriosus which permits blood to escape directly from the pulmonary artery into the aorta After the child is born the lungs expand and blood should travel through the pulmonary bed for oxygenation the short circuiting action of the ductus arteriosus is no longer necessary Under normal conditions this vessel becomes closed off soon after birth but in some individuals this obliteration is delayed for weeks months or even longer Christie[4] studied subjects from routine autopsies and found that the ductus was obliterated in 95 per cent of them by the end of the twelfth week in 99 per cent by the end of the first year

The mechanism of normal closure of the ductus is not fully understood but certain factors are believed to play a role in its obliteration Some smooth muscle fibres have been described in the wall of the vessel which lend support to the theory that a reflex mechanism presumably working through a pathway in the vagus nerve has something to do with diminution in the size of the vessel —even though this might not account for its full closure Evidence for such a mechanism of closure was obtained by the fascinating observations of Barclay Barcroft Barron and Franklin[3] It is possible that the chemical constituents of the blood also influence contraction of the ductus wall as was demonstrated by the experiments of Kennedy and Clark[20] A third mechanism in ductus closure is dependent upon the change which normally occurs in spacial relationships of various mediastinal structures after a child is born When the lungs expand the pulmonary artery assumes a different position with respect to the aortic arch and it is obvious that such a shift will angulate the ductus which lies between them While all of these factors probably play a role in diminishing the size of the ductus the ultimate obliteration of the vessel depends upon degenerative changes within its wall the histologic sequences of which have been frequently commented upon

It is important to note that the normal closure of the ductus

arteriosus is not accompanied by thrombosis of this channel Indeed the appearance of a clot within the lumen of a ductus or the formation of a thrombus on either end of a closed ductus must be regarded as a pathologic process The dangers of such a thrombosis are at once obvious, since these clots may become a focus from which emboli are thrown off into either the pulmonary circuit or into most parts of the peripheral arterial circulation Such embolic phenomena may be observed in later life but in most instances they occur within the first month of life Under such circumstances the closed or thrombosed ductus will not produce a murmur There may be evidence of infarcts in the lungs but more commonly arterial embolism produces ischemia or infarction in the brain various abdominal viscera, the kidneys and particularly in the legs Clinical and pathological observations from such patients have been previously commented upon [1]

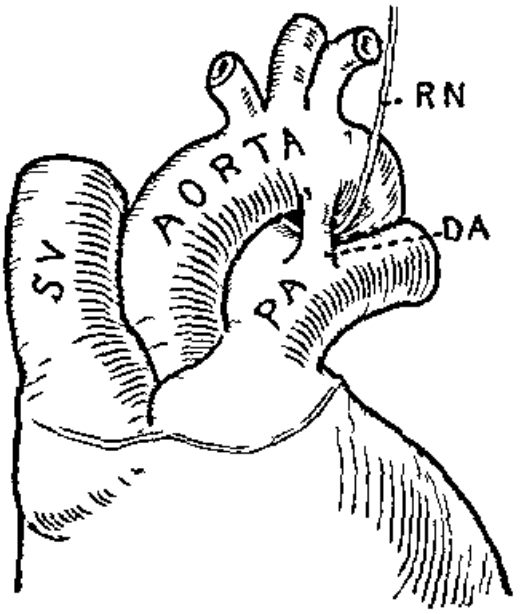


Fig 1 Sketch of great vessels indicating position of a patent ductus arteriosus between the aortic arch and pulmonary artery The left recurrent laryngeal nerve, RN, courses around the aortic arch lateral and posterior to the ductus

When the ductus arteriosus does not close and remains open beyond the first year or two of life, the individual is left with a shunt which is essentially an arterio venous fistula After birth, the direction of blood flow in the ductus becomes reversed because the pressure within the aortic arch is now higher than that in the pulmonary artery, hence, blood now passes from the aortic arch into the pulmonary circuit Such a communication may be tolerated extremely well if the individual is fortunate enough to escape any superimposed infection, and if the ductal shunt is a relatively small one Under such circumstances, individuals have been found to have little or no incapacitation and have lived to advanced years However, such a fortunate outcome is not in store for most individuals who possess a patent ductus arteriosus There are certain hazards which are well recognized and which occur rather fre

quently (1) The shunt may divert so much blood from the aorta that the peripheral circulation is deficient and the individual has a retarded physical development While such subjects may be below par in weight and height as a rule they have normal mental development and capacities (2) The heart may increase its output in an attempt to maintain the peripheral circulation at a satisfactory level but in doing so an extraordinarily large amount of blood is shunted through the ductus Under such circumstances the individual may be relatively well developed and indeed be entirely normal in appearance and yet there is evidence of cardiac embarrassment or failure (3) There may be superimposed upon this abnormality a bacterial infection usually with *streptococcus viridans* organisms The frequency with which bacterial infection occurs is difficult to estimate with any accuracy It is reasonable to believe that it is found in about twenty five per cent of individuals who live well into adult years (4) There are more rare complications such as aneurysmal dilatation and rupture The first of the above named complications appears in childhood, whereas the others are more apt to be problems of adult life particularly in the third and fourth decades

To date the best studies on the prognosis for individuals with an untreated patent ductus arteriosus have been made by Keyes and Shapiro[2] They point out that patients who are alive at seventeen years of age with an open ductus have a life expectancy which averages about half that of the population as a whole It is apparent that the patent ductus arteriosus often seems to be a benign abnormality when viewed in early life but long term follow ups show that the outlook is serious both from the possibility of ultimate incapacitation and from the shortening of life which are apt to be brought about by the malformation It is this general picture which has given an impetus for the search for surgical methods of closure of the vessel in the hope of relieving complications which have already appeared and also in avoiding others which have not yet occurred

Diagnosis

If one consults older textbooks of medicine many statements which we now feel to be erroneous are found regarding the symptomatology and the recognition of a patent ductus arteriosus Since nothing in a therapeutic way was available at those times it was

the general custom of physicians to be satisfied with classifying the malformation as "congenital cardiac disease" Now that the cardiovascular abnormality can be treated by surgical means, greater attention should be paid to the symptomatology and physical findings in individuals with the lesion Fortunately, it is possible to recognize the condition with a high degree of accuracy

In recent years attention has been focused by Gregg[10] and others on the incidence of rubella in a mother, during the first trimester of pregnancy, and the association of congenital abnormalities in the fetus resulting from such a gestation In two instances I have known of such infection in the mothers with appearance of a patent ductus arteriosus and congenital cataract in the children These represent only about one per cent of individuals which have been personally observed with an open ductus While some cardiac defects may be reasonably explained on the basis of an arresting disease which occurred early in fetal life, it is fair to assume that a persistence of the ductus arteriosus will not be found to have any such etiological basis Indeed, a patent ductus arteriosus does not represent a fetal abnormality of any sort, instead, it is a failure of normal closure *after* the child has been born

Patients with a patent ductus arteriosus may have little or no evidence of cardiac embarrassment or they may have marked cardiac invalidism, depending upon the age of the individual and the size of the leak which exists In general, the abnormality is well tolerated in childhood years and frank decompensation is rare in that period A youngster can have boundless energy, indulge in strenuous exercise, and may appear to be entirely normal to its parents More frequently, there is slight to moderate limitation of physical activity, and it is evident that excessive exercise is poorly tolerated or is followed by dyspnea, palpitation, or undue fatigue Patients in mid life often have moderate embarrassment, less commonly they may have actual failure Often the adult is conscious of the fact that he, or she, cannot maintain former levels of work, that fatigue is excessive or that long periods of rest must be taken in order to carry on with a reasonably active life I am becoming increasingly impressed with individuals who present themselves in the thirties or forties, who have no frank symptoms or signs of cardiac failure but who have lost their pep and who drag on their daily existence with no exuberance While such people are not bedridden, nor are they invalids in the common sense of the term, they are nevertheless incapacitated and are limited in their ef-

ficiency and usefulness because the heart is overburdened by an excessive load which an open ductus places upon it

The general physical development of the individual may be somewhat retarded a finding in an appreciable number of cases When compared to normal children the height and particularly the weight are apt to be less than the average normal and in some instances these findings are striking In many cases the physical growth has not been impaired and indeed it appears to be unusually good

When streptococcus viridans infections have become superimposed upon an open ductus certain points are in evidence It is rare for such infection to be found in childhood though we have seen it in a girl of four years The highest incidence of endocarditis or pulmonary endarteritis is found in the third or fourth decades The complaints include fever excessive sweating weight loss anorexia hemoptysis or chest pain (from pulmonary infarction) or changes in various parts of the body suggesting arterial embolism (from vegetations which develop on the mitral and aortic valve in the later stages of the disease) Petechiae or ecchymoses of the mucous membranes or skin should certainly suggest the correct diagnosis Blood cultures prove the presence of this complication and probably give some evidence regarding its severity

The physical findings in an uncomplicated case of patent ductus arteriosus include certain features Associated defects in other parts of the body are rare The color of the skin and mucous membranes is normal in most instances but some pallor is present in others Cyanosis is never found unless the individual has frank cardiac failure There is no clubbing of the nails The heart may be of normal or slightly increased size great enlargements are quite rare The activity of the heart may be within normal limits but if the ductus is large the cardiac impulse has an increased forcefulness and a heaving pulsation is transmitted up into the neck vessels On auscultation a very characteristic murmur is heard in the pulmonic region that is the second and third interspaces to the left of the sternum It is continuous accentuated during systole and dies off during diastole It usually has a very rumbling quality which distinguishes it from other cardiac murmurs It has been described as a machinery murmur and the clinician who has listened to several of these patients should certainly be able to identify the murmur thereafter The murmur may be widely transmitted over the precordium into the left axilla up

into the neck or over the back—particularly to the left of the spine. While all of the murmur may be transmitted it is more common to have only the louder systolic element carried to the cardiac apex, the neck vessels or to the back. In general a ductus murmur is one of considerable intensity and is accompanied by a thrill in about half of the cases. This thrill may be continuous or it may be limited to systole. It is most intense over the pulmonic region and is usually not transmitted far beyond this area. On theoretical grounds it is possible for some patients who have a tiny ductus to have a murmur which is limited to systole yet from a practical point of view a murmur that is limited to systole almost always represents some other cardiac abnormality.

The blood pressure usually shows a systolic level which is essentially normal for the age of the individual. In two adults we found some degree of hypertension, a fact which was thought to be related to other pathologic processes and not to the ductus itself. The diastolic level will be normal or suppressed depending upon the size of the ductus. Smaller fistulae do not give any important change in the diastolic pressure, but if the leak through the ductus is great the diastolic level will be strikingly diminished to 50 or 40 millimeters of mercury. When the pulse pressure is high there may be a Durosier's sign or a visible capillary pulsation in the nail beds.

Laboratory data are always within normal limits. These patients do not develop a polycythemia.

Electrocardiographic tracings are helpful particularly from the negative evidence which they generally give. Fibrillation or other indications of myocardial damage may be found in older subjects when the strain on the heart has been excessive. In most cases electrocardiograms are normal and there is no axis deviation. In a few tracings we have seen some left axis shift particularly in older individuals who exhibited definite cardiac embarrassment. In no instance have we found a right axis deviation. This is a point of extreme importance since the detection of a right preponderance should make one suspect the presence of some other lesion particularly a pulmonic stenosis. A prolonged PR interval would suggest that the auriculoventricular conduction apparatus is longer than normal and may be stretched out around an interventricular septal defect.

Roentgenologic studies may help in the recognition of a patent ductus arteriosus but they are also an aid in ruling out other

Fig. 2.—Heart film from a patient with a proved patent ductus arteriosus. The heart is slightly enlarged, the pulmonary artery (indicated by arrows) is a little more prominent than normal, and there is slight fullness of some vessels of the lung fields.

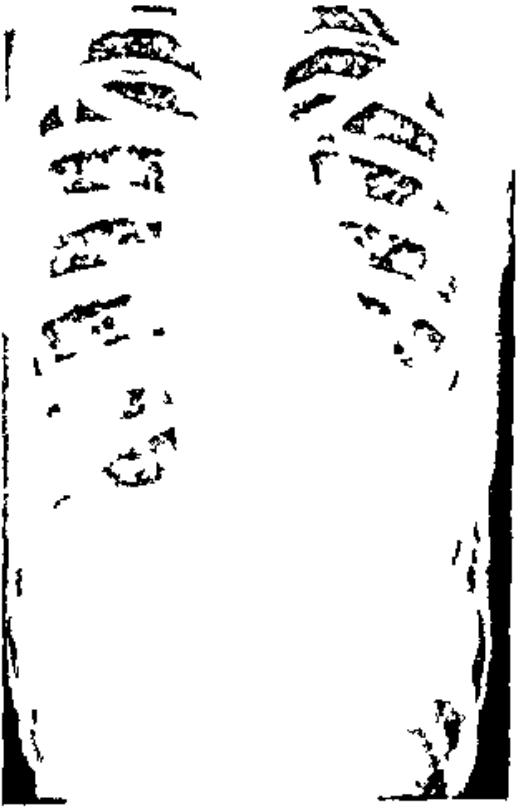


Fig. 3.—Roentgenogram of a fifteen-year-old girl with severe cardiac disability from a large patent ductus arteriosus. The heart is moderately enlarged; there is no definite fullness of the pulmonary artery, but there is some vascular congestion in the lungs. Note the long, narrow chest and the extreme thinness of the soft tissues outside of the thorax.

cardiac abnormalities or rheumatic valvular disease. In general, when the ductus is small the roentgenological picture may be normal or show little change therefrom. Conversely, when the ductus is of moderate or large size there are certain findings which are obvious in the film or fluoroscopic studies (Figures 2, 3, and 4). The heart is slightly or moderately enlarged, particularly in its transverse dimensions. Marked enlargements are rare. While it may be difficult to tell whether one or both ventricles are hypertrophied, not infrequently is it impossible to show that the left chamber is predominant. Since blood is flowing in increased volume into the pulmonary artery, this vessel (frequently incorrectly called the pulmonary conus) is fuller than normal and projects outward from the upper left border of the cardiac shadow. Likewise the vessels within the lungs, particularly around the hila, are apt to have an increased fullness and prominence. In some instances the hilar vessels may be found to have a hilar dance, which is not a transmitted impulse, but which is an intrinsic increased amplitude of pulsation of these arterial branches. This is quite difficult to ob

serve and too much reliance should not be placed upon the presence or absence of this point Left anterior oblique and lateral views give evidence of left auricular enlargement in about one half of the cases This dilatation is best seen by an encroachment on the barium filled esophagus Enlargement of the left auricle commonly seen in patients with mitral stenosis of rheumatic origin can also appear from an uncomplicated patent ductus arteriosus This is dependent upon the increased blood flow through the left side of the heart Fluoroscopic observations or kymographic tracings generally show a heart with an increased amplitude of pulsation particularly over the region of the left ventricle but likewise in the aortic knob and in the pulmonary artery

Fig 4 Right anterior oblique film from a patient with a patent ductus arteriosus indicating the posterior enlargement of the left auricle (shown by arrows) This is a frequent finding in these patients

A patent ductus arteriosus is not difficult to detect it can be recognized in over ninety five per cent of the cases with great facility While electrocardiographic studies and roentgenologic observations are important, it is well to emphasize that in the vast

majority of cases a few minutes of intelligent auscultation with the stethoscope is the prime factor in the recognition of this condition Furthermore if a characteristic murmur does not exist in a given patient too much stress should not be laid on laboratory or ro

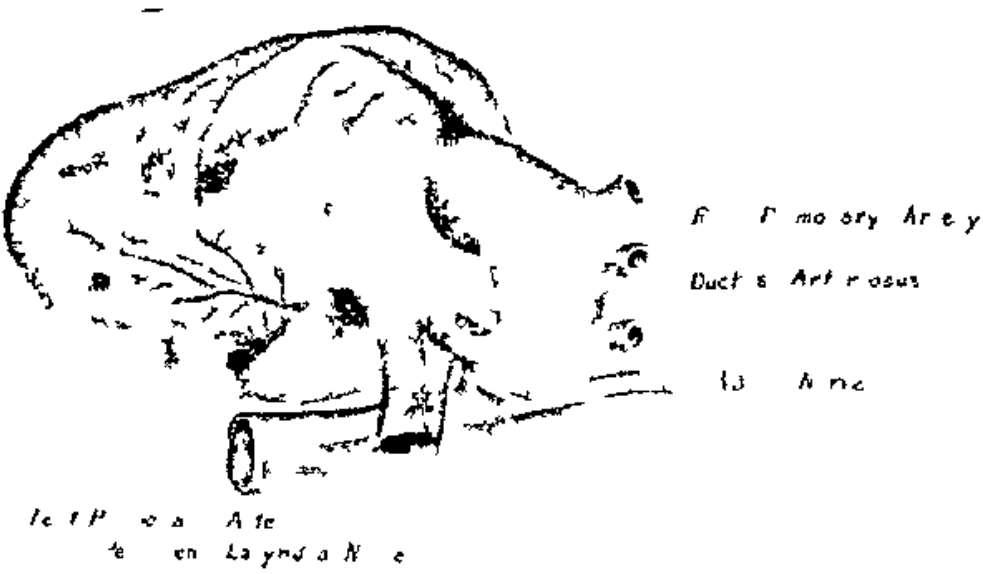


Fig 5 Drawing of the heart and great vessels in a view from which they would be seen by the surgeon operating through a left antero lateral thoracic approach

cutgenological findings which suggest the presence of a ductus because operation under such circumstances will almost certainly lead to the finding of some other congenital cardio vascular defect

Selection of Cases for Operation

As work has progressed with the surgical therapy for the patent ductus arteriosus ideas have changed regarding the selection of patients for operation There is general agreement that certain individuals should be operated upon whereas there is still some debate regarding the desirability of operation in others Certainly the child or adolescent who is not developing properly in physical stature will derive great benefit from closure of the shunt Similarly, the individual who has some evidence of cardiac embarrassment or failure can have his burden greatly reduced by ductal closure The patient with subacute bacterial endarteritis or endocarditis can likewise be helped in the majority of cases by closure of the shunt Whether or not surgery should be employed in preference to penicillin therapy for infected cases is now open to ques

tion since approximately the same percentage of permanent cures can be obtained by either method

Many patients are seen in the first part of life when they are relatively free of symptoms and the problem arises regarding the desirability of operating in the hope of avoiding future complications It was my original contention that operation was not justified under such circumstances, but this view has now been altered The mortality rate for operation on the ductus in children in the present series is exceedingly low and is about one per cent This *negligible risk is far less than the risk of letting these individuals* go untreated Hence I have adopted the policy of advising surgery for all individuals in the childhood period who have a patent ductus *arteriosus even though they are symptom free at the moment* This policy is adopted mainly for two reasons First an increasing number of individuals are presenting themselves in mid life with serious *fatigue impairment of general efficiency or actual failure and a* review of the histories indicates that they had no complaints in early life Second there is a tremendous difference in the technical *difficulties encountered in the childhood period as compared with* those in adult life Before puberty one can practically guarantee that a permanent and complete closure of the ductus can be effected *In contrast the adult presents certain features which* greatly complicate the undertaking and hence tax the surgeon's ingenuity In adults it is more difficult to get a satisfactory exposure and the great vessels are apt to be adherent to one another so that their separation is fraught with the risk of serious hemorrhage The ductus itself and the great vessels are much more rigid will not stand as much manipulation and are more subject to serious injury With increasing years the ductus tends to become shorter and there is less room to work upon it For these reasons I have great enthusiasm for operations on young subjects before they develop complications and I have considerable hesitancy about operation in some of the older patients when complications have set in

In summary I do not believe that these procedures should be undertaken in adults unless there are very clear indications for the necessity of operation because the risk of nonsurgical therapy is probably lower than the risk of surgical attack In contrast the *results have been so satisfactory for operations in the childhood* years that a rather widespread use of this procedure is justifiable

in the hope of avoiding future difficulties This policy should be adopted only by those who are able to demonstrate a reasonably low mortality rate in an extended series of cases The widespread use of this procedure by occasional operators with a limited experience in this field is not justified because performance of the operation by such individuals might inflict higher mortality rates upon patients than would be the case if they were left without surgical therapy

A word of caution is necessary regarding those individuals who have any cyanosis and also the typical signs of patent ductus arteriosus Under such circumstances the ductus should never be closed since it is almost certainly acting as a compensatory mechanism for some other cardiac defect

An interventricular septal defect or a rheumatic mitral stenosis is not a contraindication to operative closure of the ductus—since the ductus does not compensate in any way for these other lesions Obviously operation on such patients will not restore the heart to normal but at least the organ can be improved and can be relieved of some of its strain

Surgical Technique

The operative approach in all cases of the present series has been through a left antero lateral incision traversing the pleural cavity temporarily collapsing the lung and viewing the mediastinum from its lateral aspect (Figure 6) If the thoracic wall is properly opened it gives an admirable exposure The cutaneous wound was originally made above the breast but in all recent cases it has been made in a curvilinear fashion below the breast extending from the edge of the sternum downward and outward into the axilla almost to the posterior axillary line This opening is developed down to the pectoral fascia and the entire breast is turned upward and outward This is a particularly important step in a woman because failure to mobilize the breast off of the pectoral fascia will greatly limit the subsequent exposure This extensive elevation of the breast has in no way interfered with its blood supply subsequent appearance et cetera The pectoral major and minor muscles are cut across and are detached from the chest wall The thorax is entered in the third intercostal space The third and second costal cartilages are cut so that the ribs can be pushed upward and held in that position by a self retaining retractor The lateral and posterior portions of the third intercostal muscles are

now severed almost around to the angle of the ribs The anterior border of the latissimus dorsi muscle is severed and the anterior serratus muscles are divided backward as far as the long thoracic nerve All of these steps are necessary to allow full spreading of the wound

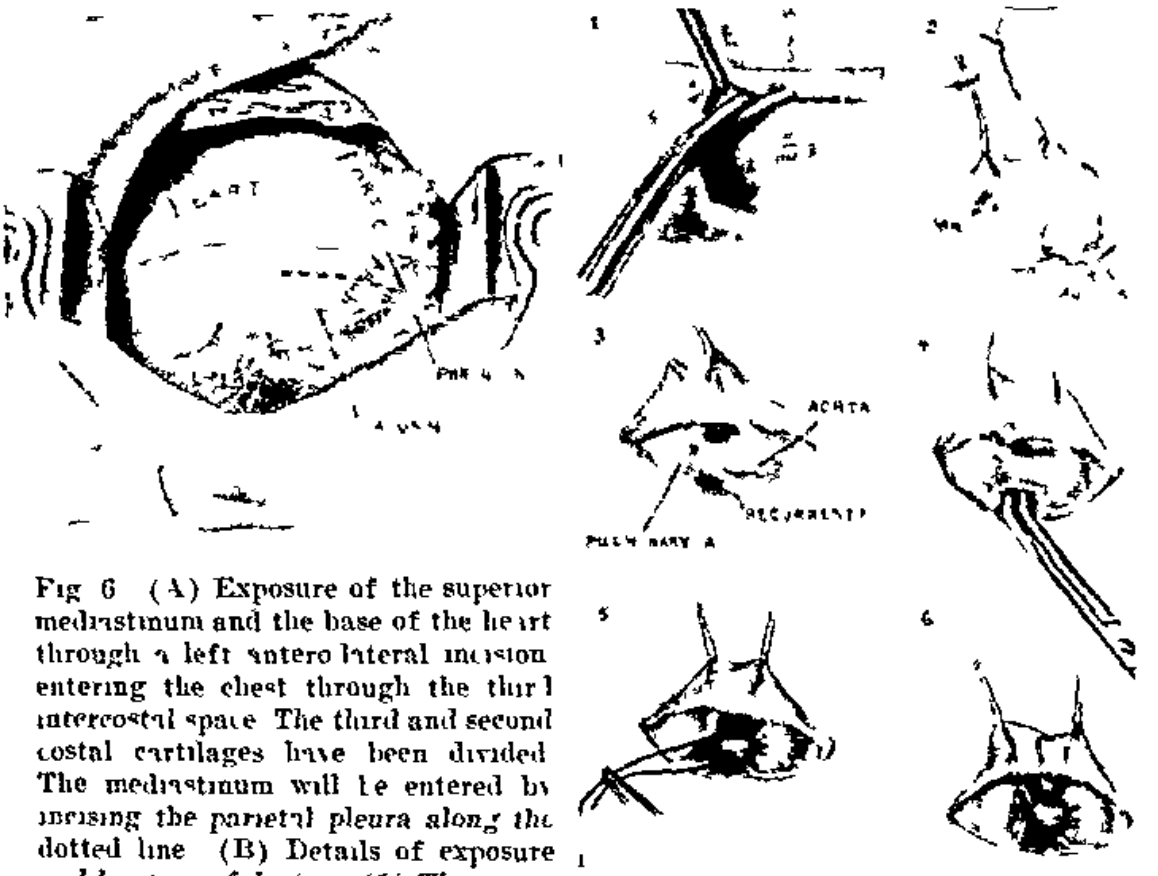


Fig 6 (A) Exposure of the superior mediastinum and the base of the heart through a left antero lateral incision entering the chest through the third intercostal space The third and second costal cartilages have been divided The mediastinum will be entered by incising the *parietal pleura along the* dotted line (B) Details of exposure and ligation of ductus (1) The parietal pleura of the mediastinum is being opened over the aortic arch about halfway between the phrenic and vagus nerves (2) The anterior flap of pleura is *held forward with one or two sutures* Within the *underlying fat and areolar* tissue the vagus and recurrent laryngeal nerve are identified (3) Preventing portions of the ductus, aorta, and pulmonary artery are cleared of fat and overlying tissues (4) The back wall of the ductus is being separated from the mediastinal structures by blunt dissection (5) The ductus has been freed and *one heavy braided silk ligature* has been placed and tied *The second ligature* is being brought into place (6) Two ligatures have been used, leaving several millimeters of tissue between them

Palpation of the mediastinum reveals a continuous thrill which can be felt widely over the great vessels but which is most intense over the ductus and in the pulmonary artery adjacent to it The parietal pleura is now opened parallel to and behind the phrenic nerve (*Figure 6, continued*) *One then comes down upon fat,* areolar tissue, and a group of lymph nodes which must be cleared

way to view the underlying structures It is well to identify the left vagus nerve as well as the recurrent nerve which comes from it and courses around under the aortic arch posterior to the ductus In no instance of the present series has there been any operative injury to these nerves Sometimes it is necessary to divide branches

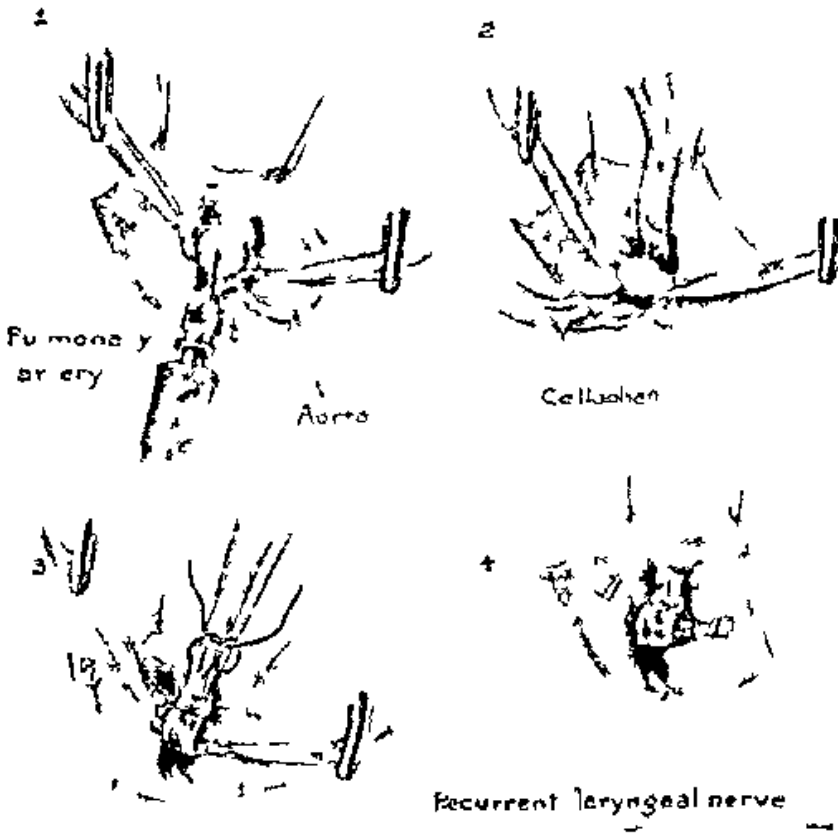


Fig Technique formerly employed for wrapping ductus with cellophane to produce regional sclerosis (1) Ductus has been freed and two linen umbilical tapes have been placed Sclerosing fluid was injected in a few instances (2) Cellophane brought into place A sheet of cellophane about one inch wide has been folded on itself four times to make a strip about a quarter of an inch wide and of four thicknesses (3) Cellophane tied on ductus (4) The end of cellophane is tied with a fine silk ligature to make sure that it will not come untied The ends of the linen tape are likewise tied or are held together with a silver clip

of the vagus nerve which run down to the left lung root in order to gain adequate room in this region No post operative deleterious effects have been observed from such divisions As the dissection is carefully continued the posterior and anterior borders of the vessel can be defined Deeper dissection can then be started between the aorta and the adjacent pulmonary artery The exploration posterior to the ductus is at first somewhat blind and must be performed with extreme care and with blunt instruments While

working behind the ductus it is well to keep the dissection upward toward the aortic arch and, hence, away from the thin walled pulmonary artery which is less tough and less able to withstand manipulation and mechanical injury As the operator proceeds, enough room can be obtained between the aortic arch and the pulmonary artery so that the back of the ductus can be viewed and a clear space can be seen between the ductus and the underlying left main bronchus It is of great importance that this dissection be adequate and thorough Negligence in the performance of this step accounts for many of the failures which have been reported following ligation of the ductus If the vessel is ligated without adequately freeing it, the chances for erosion of the ductus by ligatures are greatly increased Touroff[28] has stressed the need for meticulous clearing of the structures, and we are in accord with his admonitions

The left lateral wall of the ductus must now be completely freed of the lappet of pericardium which almost always extends up over it This can be cut away from the ductus and can be pushed off of it and off of the adjacent pulmonary artery It is well to accomplish this step without opening of the pericardium, a mishap which allows frothy pericardial fluid to run down and obscure the field

Methods of closure of a ductus have undergone several stages of evolution In the earlier phases, ligation alone was employed In a few of these a single, heavy, braided silk tie was used In most instances two ligatures were applied Linen umbilical tape seemed to have only slight advantage over heavy silk ties In twenty eight patients the ductus was wrapped with cellophane in the hope that the sclerosing action which this material produces[26] would excite regional fibrosis and close off any small opening which might have been left (Figure 7) In several cases this gave a final closure within three or four months after operation, but it could not be relied upon to attain this goal uniformly In a total of 130 surgically treated cases, forty seven individuals had one of the above described types of ligation Follow up observations indicated that in about eighty per cent of the patients a complete obliteration and closure of the shunt was obtained In about ten per cent of the cases the ligature cut through and some of the fistula was re established In the remaining ten per cent of the cases, the ligatures were not put on tightly enough to close the vessel completely While these overall results were rather good, an attempt was made to find

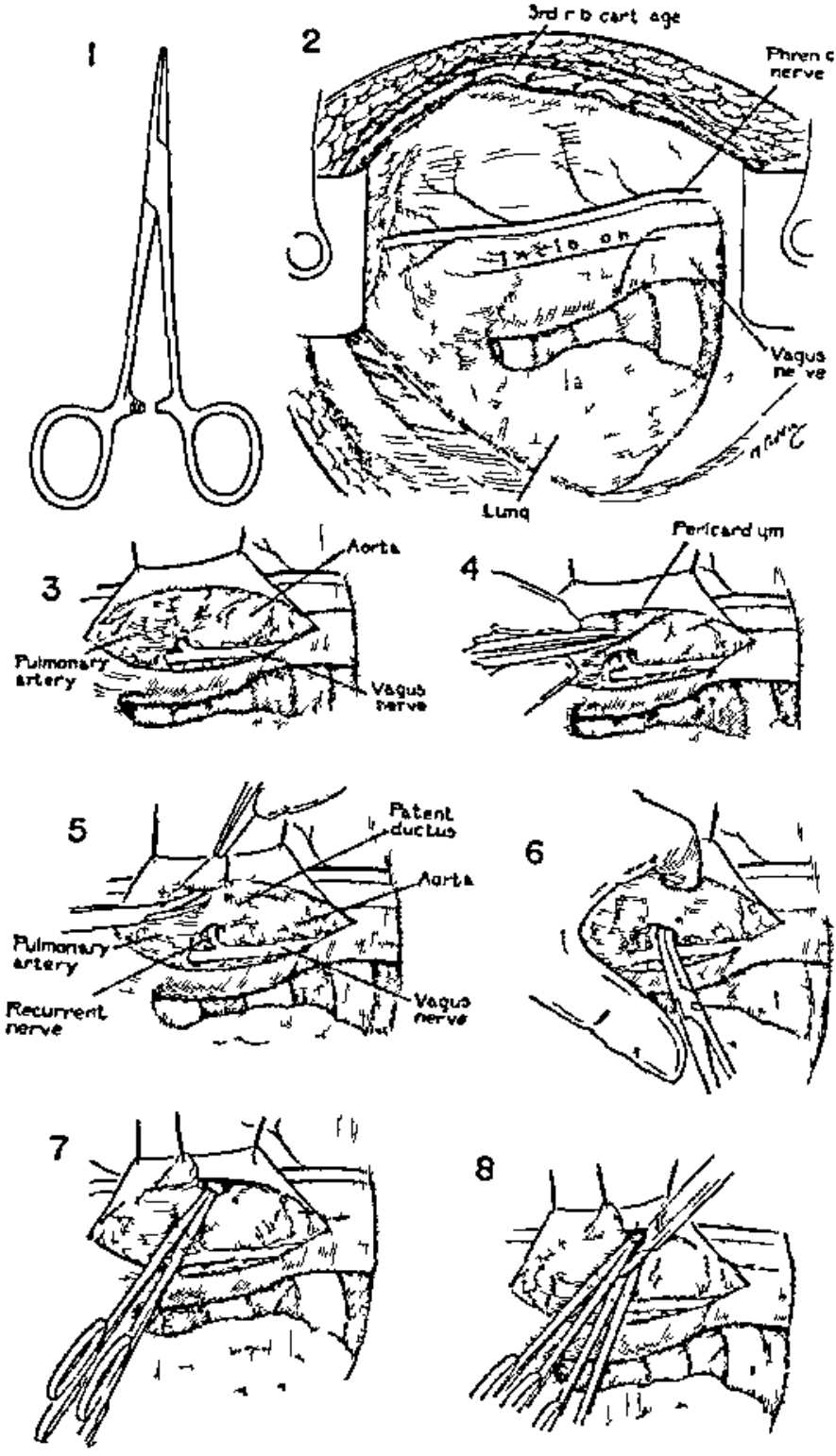
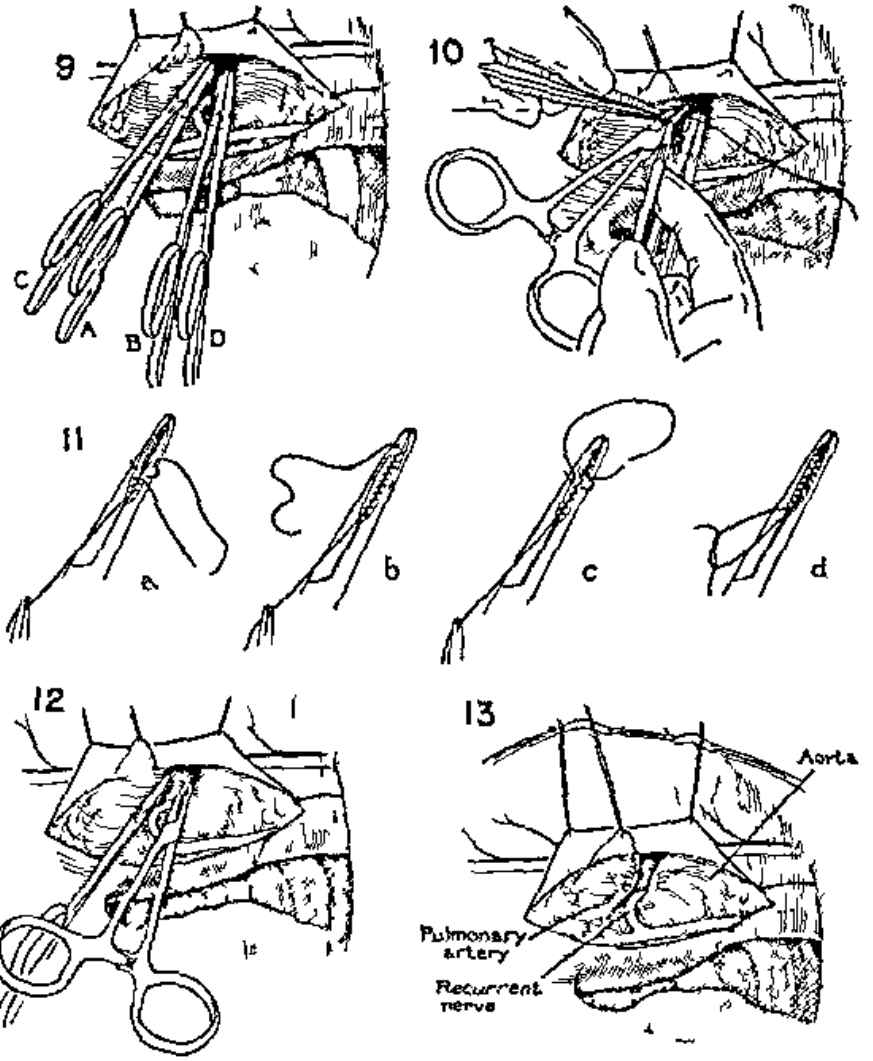


Fig. 8. Technique for complete division of the ductus arteriosus. (1) Type of clamp specially prepared for subsequent application to the ductus. This is a Crile hemostat, the jaws of which have been ground to make them a little thinner than a stock instrument. (2) Exposure of superior mediastinum through a left anterolateral approach entering the chest in the third interspace. The third and second costal cartilages have been divided. The mediastinum will



entered along the black line posterior to the phrenic nerve (3) Anterior flap of parietal pleura is held forward, exposing the underlying aorta and pulmonary artery (4) A lappet of pericardium is being identified (5) Lappet of pericardium being raised and turned caudally by sharp and blunt dissection from the underlying ductus and regional pulmonary artery (6) Anterior surface of the ductus has been completely freed Posterior wall is being bluntly dissected from subjacent structures (7) Two clamps placed upon the ductus (8) A third clamp has been squeezed on the ductus, this rides up on the pulmonary artery A fourth clamp has been placed on the opposite end of the ductus, and this rides up onto the aorta The ductus is being severed between the two middle clamps (9) Ductus completely severed, leaving two clamps on either end of the vessel (10) Pulmonary end being sutured, after removal of the presenting clamp This leaves a small cuff of tissue which can be sewed over and over with a fine silk stitch (11) Details of closure of pulmonary end of ductus The cuff of ductus tissue is being whipped over and over with a continuous 5 0 Deknatel silk stitch carried on an atraumatic needle Individual bites go through the full thickness of each side of ductus wall (12) Pulmonary end has been closed Attention now turned to aortic side from which the presenting clamp has been removed and a small cuff of ductus provided This cuff will be sewed with the same technique as was employed for the pulmonary side (13) Hemostatic clamps removed from pulmonary and aortic sides, showing individual closure of these two vessels

some other method whereby the ductus could be completely divided in all cases With these thoughts in mind the following technique (Figure 8) was adopted and has now been employed in eighty three patients with complete satisfaction

Four Crile hemostats have been prepared by grinding their blades so that they are about two thirds the thickness of stock instruments After the ductus has been *adequately* and *widely* freed two such clamps are placed upon it This usually takes up all of the available room between the aorta and the pulmonary artery However it is possible to squeeze on a third clamp which rides up somewhat on the pulmonary artery Likewise a fourth clamp then can be crowded onto the aortic end and this clamp generally rides up on the adjacent aorta In the first eighteen cases each clamp had fitted to its handles a rubber band the tension of which held the clamp closed This was done to avoid undue crushing of the ductus but this precaution is now known to be unnecessary In all recent cases the rubber bands have been discarded and the clamps have been closed using only the first ratchet there have been no untoward effects upon the vessel either at the time of operation or subsequent thereto With the four clamps in place the ductus is divided by passing a scalpel between the two middle instruments Thus two clamps remain on the pulmonary end and two on the aortic end of the ductus When the vessel has been cut the two ends separate because the pulmonary artery and aortic arch tend to fall away from one another

Attention is now turned to the pulmonary end of the ductus The presenting clamp is removed thus providing a tiny cuff two or three millimeters in length The remaining back clamp is steadied and supported by the first assistant The ductal cuff is sewed over and over with an interlocking continuous 5 0 Deknatel silk stitch carried on a tiny curved atraumatic needle Fifteen to twenty bites are taken each one of which traverses the entire thickness of both edges of the ductus wall In this way the entire end is completely closed

The aortic end of the ductus is now treated in a similar way The upper most clamp is taken off and the cuff thus made is sewed over and over with a continuous interlocking fine silk stitch closing the ends in a very effective manner

In the first eighteen patients treated by this technique the remaining hemostatic clamps were taken off and a second row of adventitial sutures were placed to reenforce the initial line of closure

This was done on the pulmonary artery as well as the aorta More recently this second layer of sutures has been omitted as a routine procedure It is now our custom to remove the remaining hemostatic clamp from the pulmonary artery and to jam a small pack in between the aorta and pulmonary artery for several minutes to permit clotting between the stitches When the pulmonary end is dry a pad can again be placed between the aorta and the pulmonary artery and the last clamp removed from the aortic end Ordinarily packing for a few minutes is sufficient to control any oozing In a few cases it has been necessary to take a few additional stitches to reenforce a part of the suture line where there was some pin point bleeding

This type of operation may seem to be hazardous and fraught with dangers and yet it has been performed and completed eighty three times without any fatality directly related to the division of the ductus In this group there have been two surgical deaths one from staphylococcus mediastinitis and the other from cardiac failure in a woman who was an extremely poor risk and who had had evidence of cardiac decompensation for a long period of time Complete division of the ductus insures that all of the leak is stopped and that there is no chance for reestablishment of the fistula The effectiveness of this operation and the absence of mortalities which can be attributed to it make me feel that any form of ligation of the patent ductus arteriosus is an obsolete procedure and I have now completely abandoned ligation in any form I would hasten to add that division of the ductus is a delicate and painstaking procedure and hence should not be undertaken by individuals who have not mastered the technique in the experimental laboratory

Results of Operation

One hundred thirty patients have been operated upon forty seven by ligation and eighty three by complete division of the vessel There have been five surgical deaths an overall mortality of 3 8 percent The youngest patient was eleven months the oldest forty seven years of age Those who have survived the procedure have had certain changes which will be individually considered as follows

Changes in blood pressure —Following surgical closure of the ductus there is no important change in the systolic blood pressure (Figure 9) In occasional individuals the systolic pressure will rise

ten to twenty millimeters of mercury for several days and then recede to its preoperative figure The diastolic pressure however shows a marked rise and this is evident at the operating table as soon as the ductus is closed The degree of rise will vary inversely with the depression which existed prior to operation In other

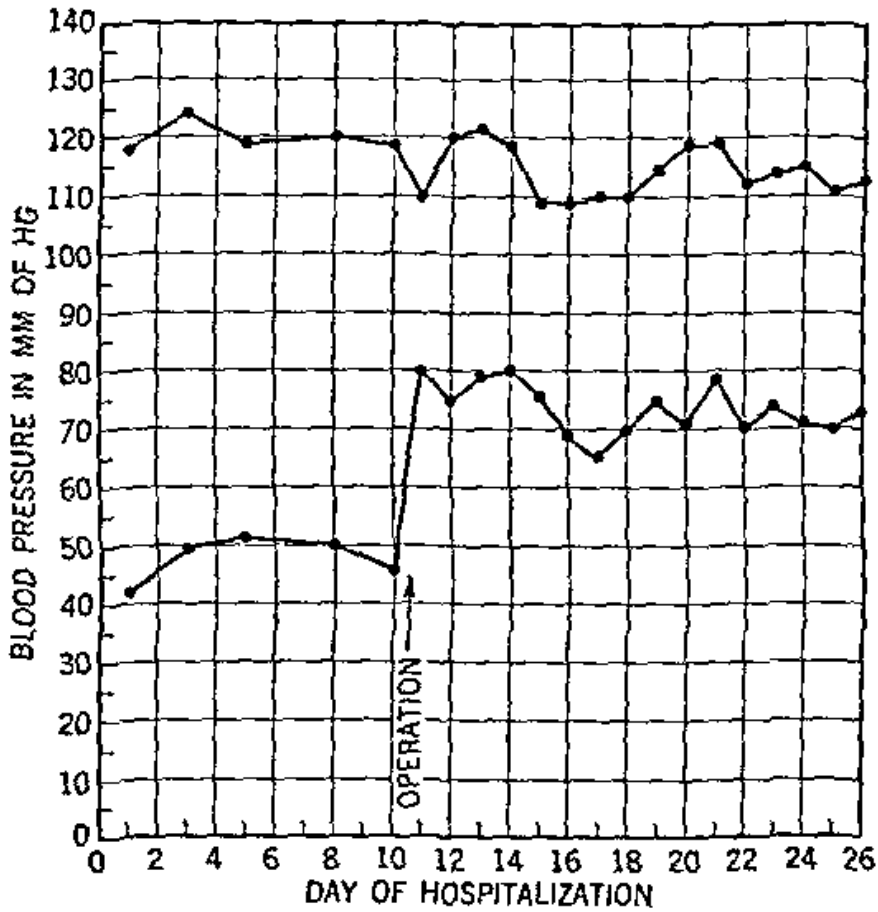


Fig 1 A typical blood pressure chart of a child before and after closure of an open ductus Operation makes no important change in the systolic level Closure of the ductus is followed by a sharp and permanent rise in the diastolic pressure

words a preoperative diastolic pressure which is not greatly below normal will change but little following operation Conversely when the diastolic pressure has been depressed to fifty forty or thirty millimeters of mercury before operation one finds an abrupt and striking rise in this reading after closure of the shunt In short when the leakage from the aortic arch has been stopped the vascular system is able to maintain the diastolic pressure at normal physiological levels

The murmur —In eight of the earlier cases where ligation was done, a murmur, of lessened intensity was found after operation

indicating some leakage through the ductus In about half of these it was believed that the ligatures had not been placed tightly enough to close the entire fistula In the other half there was no murmur in the immediate postoperative period but this was followed by reappearance of the same during the second or third postoperative week In these latter instances it was obvious that the ligatures had cut through the vessel to some degree

In five individuals all of the ductus murmur has disappeared but there remains a murmur indicative of some other associated lesion Three of these were recognized or suspected before operation one having a rheumatic mitral stenosis and insufficiency and the other two an interventricular septal defect In the fourth and fifth cases the second lesion was not suspected prior to operation because the ductus murmur was so loud that it completely overshadowed the murmur of the second abnormality

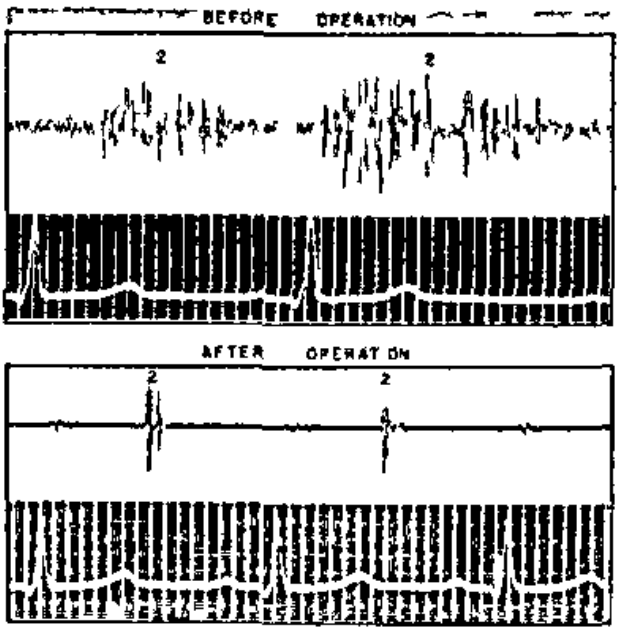


Fig 10 Sound tracings taken from the pulmonary region of the chest before and after operation (1) and (2) indicate the times when first and second cardiac sounds should occur Before operation there is a continuous murmur which is most intense during systole and which diminishes in diastole Following operation there are pure first sounds and second sounds and there is complete disappearance of the murmur

In the vast majority of cases particularly in the recent ones where complete division has been performed there is complete disappearance of murmurs after operation (Figure 10)

Activity of the heart —One of the striking changes which can be observed is the sparing effect upon the heart and the diminution in the forcefulness of the cardiac beat By inspection of the chest, particularly in a thin individual the pulsations at the cardiac apex can be seen to be less intense Likewise the pulsations over the neck vessels are less prominent A heart which before operation had a very heaving pounding and forceful beat, will be found to

have a postoperative activity which in comparison is quiet and much less vigorous Further evidence of this change can be found by fluoroscopic examination or kymographic tracings By such means one can see that the amplitude of excursion during the cardiac contraction returns to normal These postoperative diminutions in cardiac action are not great if the ductus which has been closed has been a small one In contrast when a ductus of large size has been obliterated there is a great reduction in the activity of the heart after operation

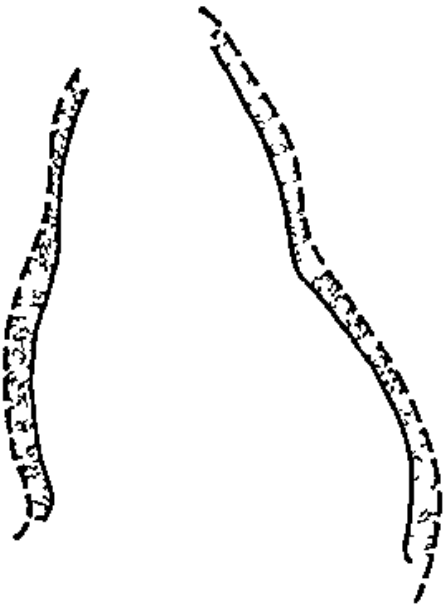


Fig 11 Changes in cardiac size and contour from an individual with surgical closure of the ductus Tracings taken from roentgenographic heart films before operation (dotted line) and after operation (solid line) Shaded portions indicate diminution in the shadow of the heart and great vessels produced by closure of the ductus

Size of the heart —It has long been known that artificial establishment of an arterio-venous communication produces cardiac enlargement of two types There may be cardiac hypertrophy and there may be cardiac dilatation Usually some degree of both exists if the shunt is kept open for a considerable period A patent ductus arteriosus is one form of arterio venous fistula and it produces similar effects upon the heart When cardiac hypertrophy has taken place the organ does not shrink following closure of the shunt However there have been observations to show that in a growing individual the thorax and other body measurements can increase whereas the heart grows very little during the ensuing year and a half or two years at the end of this time a normal cardio thoracic ratio becomes established When enlargement of a heart is primarily on the basis of dilatation the heart will shrink *very rapidly following closure of the fistula*

Measurements of the heart size can be made quite accurately with seven foot heart films before and after operation In many individuals particularly where the ductal shunt has been small there is little diminution in the size of the heart following operation In contrast when a fistula of large size has been closed the overall dimensions of the heart will shrink particularly the hori

zontal diameter (Figure 11) We have seen diminutions in transverse dimensions of as much as a centimeter and a half

Weight changes —Individuals who have an essentially normal physical development prior to operation, show no important growth changes when followed for some months or several years after operation However, underweight subjects, most of whom are in the childhood group, will exhibit a surprising and gratifying gain in

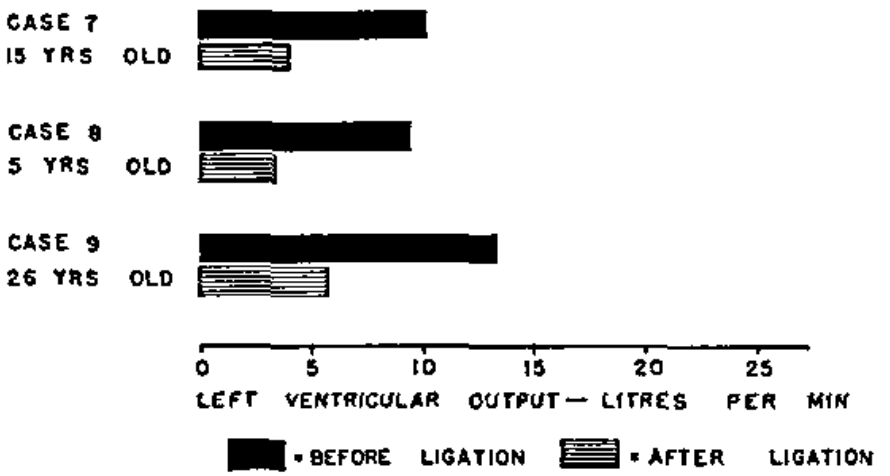


Fig 12 Graphs of data from three patients showing output of left ventricle (under conditions of operation), before and after closure of the ductus From studies by Burwell and Eppinger In each instance there is a marked diminution in the left ventricular output following closure of the ductus

weight, sometimes to an amazing degree Many of these children have added 25 to 30 per cent to their weight during the year or year and a half following operation Apparently, closure of the shunt increases the peripheral flow of blood to the body and thereby improves the physical state

Reduction of the cardiac output —Eppinger and Burwell[7] have accumulated data from some of our earlier cases to study the changes in the circulation which are produced by closure of a patent ductus arteriosus During operation samples of blood were taken from various intra thoracic vessels for determination of their oxygen contents After measuring the amount of oxygen which the patient was consuming, calculations could be made to ascertain the per minute flow through the periphery of the body, through the lungs, through the two sides of the heart, and finally, through the ductus itself Operation was then continued and the ductus was closed off Ten or fifteen minutes later, a second set of blood samples were collected and analyzed so that calculations could

again be made for the various flows in different parts of the system

From the publications of Eppinger and Burwell I would like to point out the left ventricular output of three of patients (Figure 12) The first a girl of fifteen years was pumping 10 3 litres of blood per minute from the left ventricle while the ductus was open this immediately fell to 4 litres per minute after the shunt was closed In a second child of five years with evidence of severe cardiac embarrassment the left ventricular output with the ductus open was 8 7 litres per minute whereas it fell to 3 3 litres per minute after ligation of the ductus The third patient a woman of twenty six years with marked orthopnea for three years had a left ventricular output of 14 1 litres per minute when the ductus was open in contrast to 6 3 litres per minute when the ductus was closed It should be emphasized that none of these observations were made under basal conditions They were made under circumstances of operation under anesthesia with the chest open with one lung partially collapsed et cetera However the conditions were exactly the same in the two sets of measurements except for closure of the ductus shunt The figures clearly indicate that obliteration of a patent ductus arteriosus can greatly diminish the work of the heart and presumably it can increase the cardiac reserve

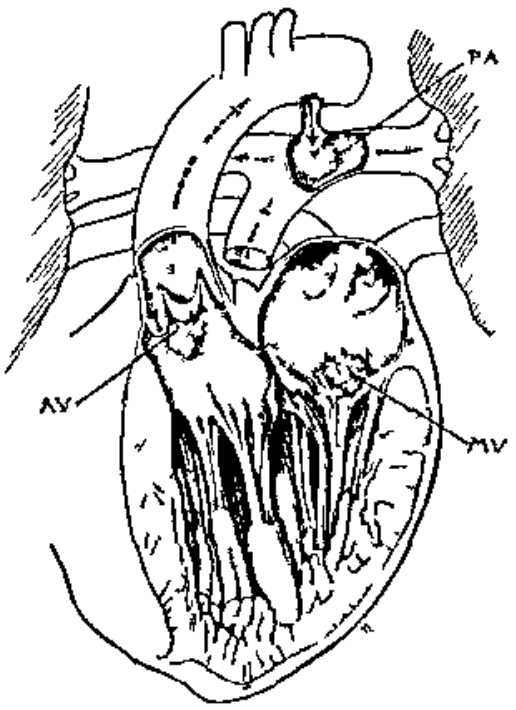


Fig 13 Diagrammatic representation of stages of bacterial infection in the presence of an open ductus arteriosus In early stages vegetations are limited to the pulmonary artery (PA) Under such circumstances closure of the ductus has very beneficial and curative effects In later stages of infection vegetations may appear on the mitral valve (MV) and on the aortic valve (AV) In such stages surgical closure of a ductus apparently will not halt the infection

Superimposed streptococcus viridans infection —Formerly I could not see any rationale for operating upon individuals with a complicating subacute bacterial endocarditis or pulmonary endarteritis It was difficult to believe that there would be any bene

ficial effects as far as the infection was concerned and indeed the risks of operation would presumably be high because of the friability of the vessel and the danger of uncontrollible hemorrhage Graybiel, Strieder, and Boyer[1] were the first to attempt operation upon an infected case but Touroff and Vesell[27] were the first to cure a streptococcus viridans infection by surgical closure of a

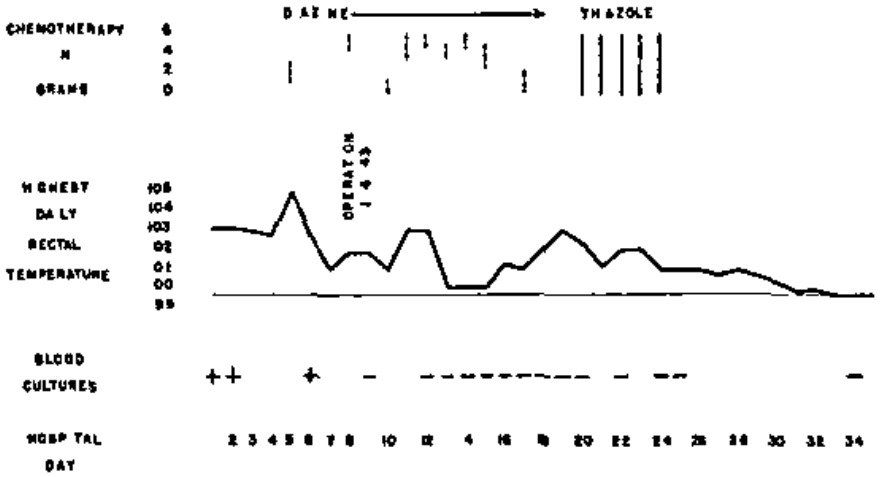


Fig 14 Clinical chart from a nineteen year old girl with a patent ductus arteriosus and superimposed streptococcus viridans infection Previous therapy with adequate sulfonamide treatment failed to sterilize the blood stream Following surgical closure of the ductus there has been subsidence of fever and all subsequent blood cultures have been negative

ductus The later reports of Touroff[29 30] show that of eleven such patients (all in the pre penicillin era) who were treated by surgery six survived and were cured of their infection Furthermore, sulfonamide therapy had been discontinued or withheld for one or another reason and the curative results could be ascribed to the operation *per se*

Experiences in our ten patients with superimposed streptococcus viridans infection closely simulate those which have been previously described by Touroff All of our patients were likewise in the pre penicillin era, but each had been treated with the sulfonamides which were available at the time In no instance could the blood stream be sterilized by chemotherapy prior to operation Sulfonamides were continued for varying periods of time after operation In three cases the infection persisted and the patients ran a downhill course, eventually dying of an overwhelming infection In the other seven individuals, the therapy was effective in bringing about a permanent cure of the streptococcus viridans infection In some of these the blood cultures became negative immediately after operation (Figure 14), while in others the cultures

remained positive for as long as two weeks after operation and then all subsequent ones were negative (Figure 15)

It is not entirely clear why surgical closure of a ductus should have curative effects upon infection which exists in the pulmonary artery, but two explanations seem plausible Blood squirting through an open ductus into the pulmonary artery must dislodge

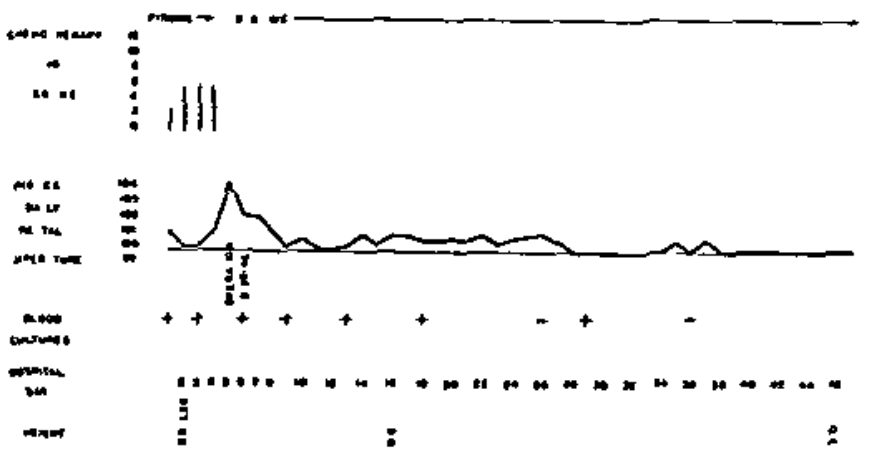


Fig 15 Chart from a thirty six year old man who for four months had shown evidence of *streptococcus viridans* blood stream infection, continued fever, great loss of weight, etc Extensive therapy with sulfapyridine and hyperpyrexia with injections of typhoid antigens had been ineffective in sterilizing the blood stream Operation as indicated In three weeks the temperature had subsided to normal, and all blood cultures since that time have been negative

bacteria from the vegetations which exist there Hence, there is constant seeding of the blood stream (Figure 13) Surgical stoppage of the flow through this nozzle will allow the current in the pulmonary artery to become more quiet and thus promote healing of the vegetations Secondly, when the ductus is open the blood within the pulmonary artery has a rather high degree of oxygen saturation, a situation which makes suitable conditions for the growth of bacteria Closure of the ductus immediately cuts off admission of arterial blood and the pulmonary artery will then contain only venous blood, the low oxygen saturation of which will discourage the growth of organisms

Touroff has pointed out that as long as the vegetations are limited to the pulmonary artery (Figure 13), operation has highly beneficial effects Such individuals may have debilitating infection, have positive blood cultures and may have pulmonary infarcts yet operation is successful in a high percentage of cases In contrast, late stages of infection usually imply that vegetations have de

veloped upon the mitral and aortic valves—a fact which can be recognized by the appearance of peripheral emboli—and under such circumstances operation has little or nothing to offer

Prior to the days of surgical treatment for patent ductus arteriosus, individuals who had superimposed streptococcus viridans infection usually died of this complication, and not more than five or ten per cent recovered under any form of therapy With the advent of surgical treatment, permanent cures have been obtained in sixty or seventy per cent of the cases, a very encouraging advance More recently, the same general results have been obtained with penicillin alone, and this type of therapy will probably supercede the surgical one While we must recognize the potency of penicillin in eradication of such infections, it is well to point out that some of these patients subsequently die of cardiac failure because of myocardial damage or exhausting effects of their illness Hence it is possible that the optimum therapy of the future will combine the antibiotic activity of penicillin with the reduction of cardiac work which can be brought about by surgical closure of the open ductus

DEFECTS OF THE PERICARDIUM

Absence of portions of the pericardium rarely gives rise to symptoms or to disturbed function of the cardiac mechanism Congenital absence of a part or whole of the pericardium has been

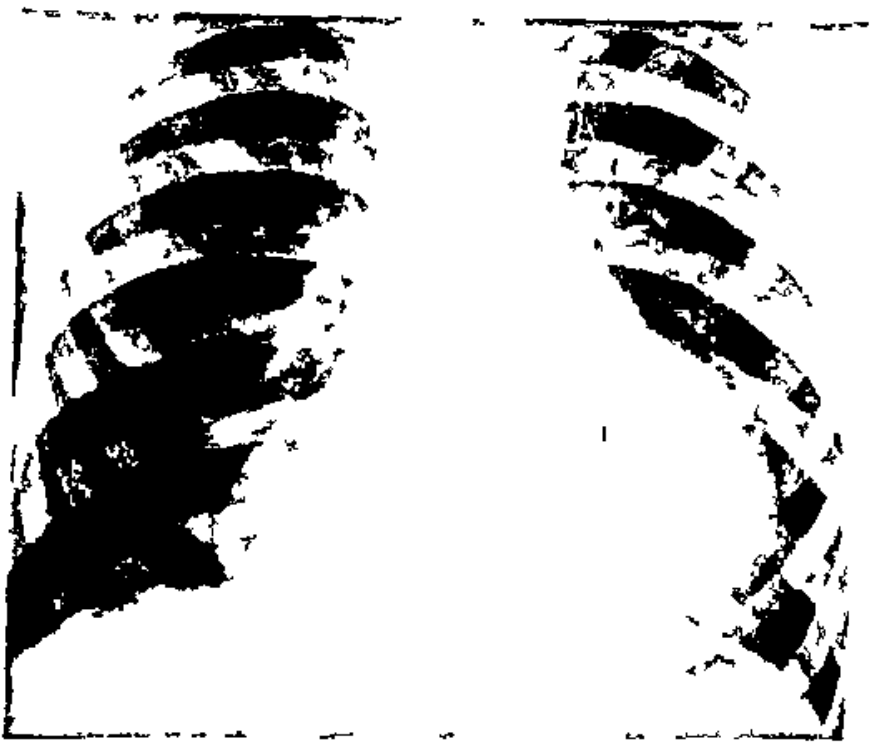


Fig 16 Roentgenogram of four year old boy with a complaint of persistent cough There is a shadow projecting from the right side of the heart (Cough was presumably caused by irritation or stretching of overlying right phrenic nerve)

observed by Ladd[24] and others these being incidental findings in thoracic operations or at the post mortem table The pericardium has been removed experimentally from animals and the adjacent structures have become smoothed over so that the moving heart can beat in a satisfactory way It is now well known that large portions

of the pericardium can be resected from humans with constrictive pericarditis and the heart can be made to move and twist in a more normal manner While absence of the pericardium, *per se*, is not deleterious, certain defects which involve the subjacent diaphragm can be troublesome, as is exemplified by the case indicated in Fig

Fig. 17 Same patient as shown in Fig 16 Visualization of interior of heart by injection of 70 per cent diodrast into the left antecubital vein Black arrows indicate iodide in right side of heart White arrows indicate mass projecting from heart This does not contain iodide, and therefore is not a dilated cardiac chamber (Operation showed this mass to be a portion of liver which had protruded through a diaphragmatic hernia into the pericardial sac)

ures 16, 17, and 18 This patient has been described in greater detail elsewhere [24]

Herniations through the diaphragm and into the pericardial sac are rare yet recognition of them is important because the individual's symptoms can be completely relieved by replacement of

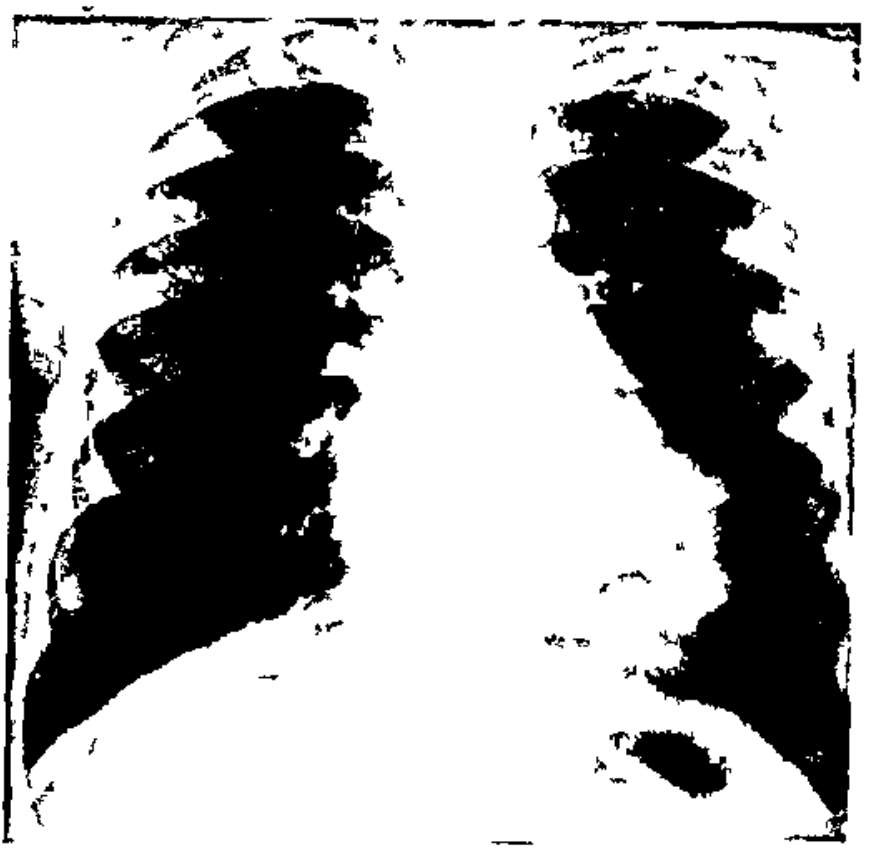


Fig 18 Same patient as Fig 16 and 17 Post operative film of heart after replacement of lobe of liver into abdomen and repair of diaphragmatic pericardial defect

the abdominal viscera into their proper position, following this with a suitable repair of the diaphragm and the pericardium

TETRALOGY OF FALLOT

A new chapter is being written in the treatment of congenital heart disease by the outstanding work of Blalock and Taussig [16] which has appeared within the last year My own experience in this field is too limited to warrant recording it here, but I would like to make a few comments on this important work, the full details of which are available in the publications of Blalock and Taussig

The description by Fallot of a combination of anatomical abnormalities serves to focus attention upon certain patients in the so called "cyanotic group" of congenital heart disease This does not include all individuals who have had cyanosis since birth, but it does apply to about three-fourths of them The clinical and pathological findings have been well described by Abbott and many other cardiologists and pathologists The studies in pathologic anatomy, which have come from various sources during the past half century, have given a broad foundation for understanding of congenital heart disease, but we have been denied therapeutic advances until recently, when a change in thinking occurred and attention became focused on the pathologic physiology of these patients Investigations such as those of Barclay, *et al*,[1] Burwell and Eppinger,[2 6 7] and others, have turned the spotlight from a mere listing of ana

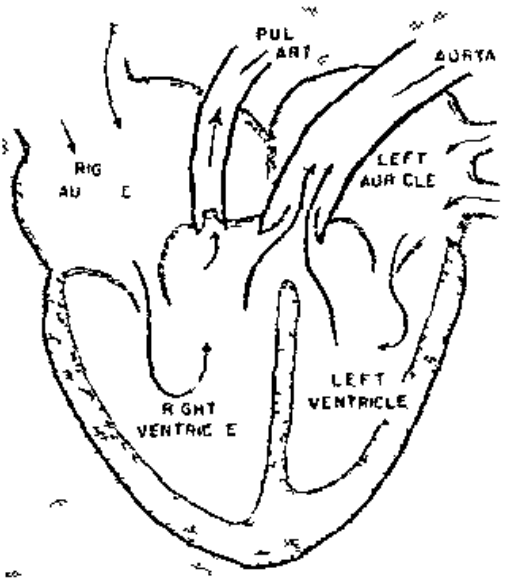


Fig 19 Sketch showing fundamental physiological abnormalities in an individual with a tetralogy of Fallot The aorta is shifted in such a way that it overrides both ventricles and receives blood from both of these chambers There is an obstruction (in some cases an atresia) at the orifice of the pulmonary artery, this may involve the valve, first portion of the artery, or the pulmonary conus below the valve The two important factors which contribute to cyanosis are (1) the direct propulsion of venous blood from the right ventricle into the aorta and (2) an obstruction in the first part of the pulmonary artery giving a diminished flow of blood into the lungs

tomical variants and have brought out into clear view what is going on in the living patient with various types of cardiac malformations With an exceedingly rich background of many years familiarity with congenital heart disease Dr Taussig has pointed out and stressed the simple fact that individuals with a tetralogy of Fallot are not getting sufficient blood through the lungs for oxygenation With this conviction she has enlisted the services of Dr Alfred Blalock who has long had a particular interest in many branches of vascular surgery The cardiologist has firmly indicated the desirability of getting more blood into the pulmonary circuit and the surgeon has devised the way for attaining this goal The results of these combined efforts are I am sure familiar to all of you and they stand as a brilliant example of what can be accomplished by the pooling of interests capacities and facilities for making a united frontal attack on a medical problem

From a physiologic view point the individual with tetralogy of Fallot has two fundamental disturbances to account for his cyanosis and the debilitating effects which are produced by the cyanotic state (Figure 19) First there is some intermixture of blood because the aorta by virtue of a dextro position of its orifice receives blood from both the left and the right ventricles Nothing can be done to correct this intermixture Second there is some stenosis of the pulmonary conus the pulmonic valve or the first portion of the pulmonary artery — any one of which diminishes the flow of blood into the lungs This physiological deficit can be overcome by the surgical establishment of a shunt between the aorta and the pulmonary artery to allow more blood to pass into the lungs The production of such a fistula is the essential feature of the Blalock operation

Blalock has devised several methods for producing a channel to increase the pulmonary blood flow (Figure 20) The left subclavian artery has been divided at the base of the neck and its proximal end turned downward and joined to the pulmonary artery Such shunts are beneficial but are frequently of insufficient size hence larger arteries have been employed such as the left common carotid or the innominate for making the fistulae In general the innominate artery anastomosis has given a shunt of appropriate size

These operations have carried mortality rates which are not prohibitive when one considers the desperate condition of many of these patients The results of operation appear to be somewhat

variable For some of these children there has been a new lease on life and the surgical accomplishment must be classed as brilliant For most of the individuals, considerable degrees of improvement *have been the rule In a relatively small number there has been a* failure It is very satisfying to see individuals who have been im

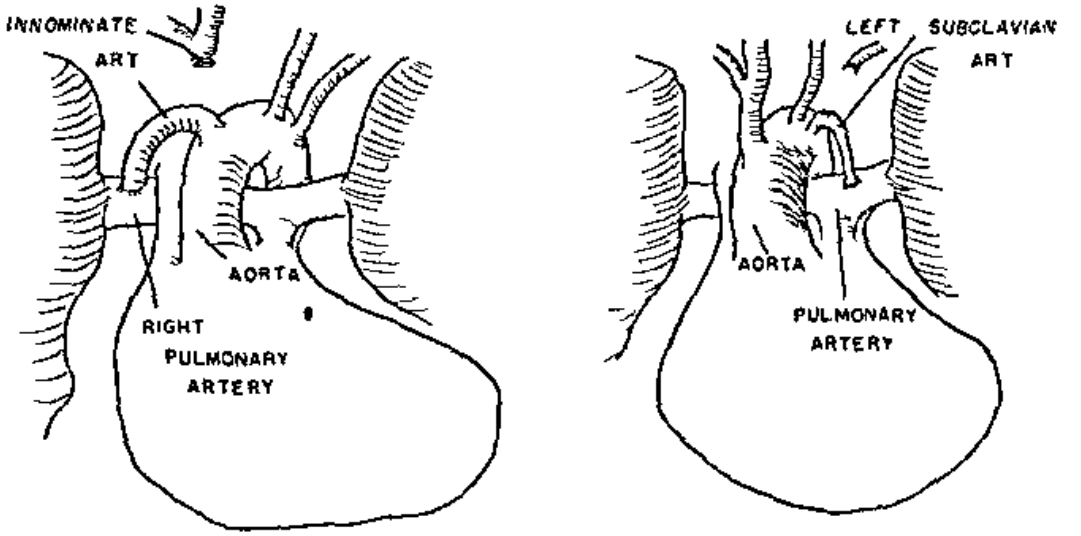


Fig 20 General techniques employed by Blalock for increasing the flow of blood into the pulmonary system *Left*—The innominate artery has been divided at the base of the neck and this vessel turned downward for anastomosis to the right pulmonary artery *Right*—The left subclavian artery has been divided at the base of the neck and turned downward so that it can be joined to the pulmonary artery The left common carotid artery has been used in some instances The techniques have usually been end to side anastomeses but in some cases a branch of the pulmonary artery has been divided and end to end unions have been made between the arterial system and the pulmonary artery of the lung root

proved by this operation The cyanosis is moderately or markedly *diminished, and objective measurements of this can be found in the* postoperative increase of the oxygen saturation of arterial blood and in the diminution of the polycythemia Furthermore individuals who have had extreme limitation of activity and who seldom did anything more than squat on the floor, can now walk or run for extended distances

Two general problems apparently need further attention and it is reasonable to assume that progress along these lines will be made in the future The first concerns itself with the selection of cases for operation At the present time the main method for picking out the individual with a small pulmonary blood flow is by *roentgenographic demonstration of a small pulmonary artery* Furthermore, there should be no evidence of pulmonary congestion It would be very desirable to have more exact methods for identify-

ing individuals with a markedly diminished pulmonary flow because it is in this group that the most promising results can be obtained by surgical means Catheterization of the right auricle and the right ventricle as well as the vessels connected with this latter chamber can yield data indicating pressures in various parts of the system and can procure blood samples from these various areas for determination of their oxygen content It is therefore possible under some circumstances to compute the pulmonary flow in an approximate manner Burwell and Dexter have made observations of this sort and I have little doubt that such examinations will be more widely used in the future It is of course, more difficult to employ the catheter technique in children but with proper sedation or possibly with general anesthesia it is quite probable that these tests can be made on subjects at younger ages than was formerly thought possible

The second problem is concerned with the development of better methods for establishing a shunt between the aorta and the pulmonary system Many of the subclavian artery anastomoses have apparently given a shunt of too small size for optimum results but the use of the left common carotid or the innominate artery has sometimes led to cerebral ischemia and a distressing hemiplegia While the neurological disorders apparently improve in time they are complications which can greatly mar an otherwise favorable outcome Blalock's observations have conclusively demonstrated the correctness of the fundamental principle that a shunt between the aorta and pulmonary artery has beneficial effects for an individual with a tetralogy of Fallot but I am not at all sure that the best method for making this shunt has yet been brought forth There is every reason to believe that another method will be developed so that the cerebral circulation can be left intact while establishing an opening of proper size between the aorta and the pulmonary artery

One cannot deny the fact that these operations make a complicated abnormality indeed somewhat more complicated There is no chance of restoring a normal cardiovascular arrangement to these individuals However the shortcomings should not in any way detract from the tremendous advances which have been made by bringing to these miserable patients a more normal appearance a greater capacity for physical exercise and also a diminution in the threat of thromboses which previously existed because of the polycythemia

RIGHT AORTIC ARCH

Apparently nothing in a surgical way has ever been attempted for treatment of a right aortic arch which gives rise to symptoms While the vast majority of such abnormalities are beyond the possibilities of surgical relief there are some circumstances under which this might be attempted In passing, I would like to record briefly several thoughts which I have had on this matter

A right aortic arch may be combined with other severe cardiac abnormalities, but in many instances it appears alone The most common form of a right aortic arch is that in which the ascending aorta projects to the right of the trachea or esophagus and then to the left behind the esophagus continuing downward as the descending aorta (not on the right side of the body but a little to the right of the normal position for a descending aorta) Great variations may occur in the large arteries which arise from the aortic arch, regarding the points of origin from the arch and also their positions as they course upward to reach the exits of the thoracic cage

Three general aspects of right aortic malformations deserve specific consideration because they raise the possibilities of surgical treatment in some patients (1) The arch may rest upon the right upper lobe bronchus and produce atelectasis or obstructive emphysema in this portion of the lung While the arch cannot be removed therefrom, it might be expedient to excise the right upper lobe if this structure becomes the seat of recurrent infection (2) In some cases, an artery arises from the arch to the right of the midline Thus a left common carotid artery or a (left) innominate artery can pass over in front of the trachea as it runs to the left apex of the chest In this way, the artery may be stretched like a bow string across the trachea and give rise to symptoms of tracheal compression The trachea could be relieved of this pressure by dividing the vessel or better still by displacing the vessel forward and anchoring it to the back of the sternum (3) The pulmonary artery, by virtue of its attachment to the aorta through the ductus arteriosus or ligamentum arteriosum, is pulled backward because of the posterior displacement of the arch Thus the pulmonary artery can be pulled against the front of the trachea in a way that compresses it There should be little difficulty in division of the ductus, or the ligamentum arteriosum, to allow the pulmonary artery to fall forward and thus give more room for the trachea and esophagus

A right aortic arch usually gives rise to few or no symptoms and the abnormality may be only of academic interest However it is possible for a number of complaints to come from these malformations as is shown by the reports of Renander [46] Sprague *et al* [48] and Gross and Ware [16] et cetera There may be dysphagia stridor dyspnea cyanosis hoarseness cough and pain in the upper portions of the chest

My attention was drawn to the therapeutic possibilities of surgery in this field by the post mortem findings in a four month old baby who died because of complications arising from one of these abnormalities This child had had difficulty in breathing and in feeding since birth Mucous often collected in the throat and gave rise to coughing but there was never any cyanosis The coughing and the collection of mucous were more troublesome when the baby was lying on her back While hesitation in swallowing had been present since birth this had become more marked in the last month of life When urged to take more than an ounce or two at a time she invariably spat up some of the formula The respirations were quite noisy and there was an inspiratory crow By x ray examination of the chest there was some peribronchitis but no other important change in the lung fields When the esophagus was visualized with a swallow of barium it was evidently pushed forward by some mass at the level of the third or fourth thoracic vertebra At this same general level the trachea was narrowed above the carina by something which pressed on its anterior surface The state of health was precarious the pulmonary infection increased and in spite of a gastrostomy which was established for feeding purposes the child succumbed At autopsy diffuse bronchopneumonia and early bilateral empyemata were found The transverse part of the aortic arch lay to the right of the trachea and esophagus and the third portion of the arch passed behind the esophagus The anterior surface of the trachea was compressed by the pulmonary artery which was drawn against it because of an attachment through a patent ductus arteriosus to the posteriorly displaced aortic arch Furthermore the left common carotid artery arose from the ascending aorta and lay tightly across the front of the trachea as it coursed upward and to the left The post mortem findings suggested that the child might have been relieved by division of the patent ductus arteriosus (to allow the pulmonary artery to fall forward) and also by dislocation of the left common carotid artery in such a way that it could be removed from the trachea

DOUBLE AORTIC ARCH

There are on record some descriptions of human subjects in whom the ascending aorta splits into two limbs, which encircle the esophagus and trachea or only the trachea, and then join to form the descending aorta In the normal development of the aortic arch, only the left fourth branchial artery persists to form the definitive arterial system [50] If, however, both the right and left branchial arteries persist, then the abnormality under consideration becomes established This malformation has been variously called "double aortic arch," "bifid aortic arch," "split aortic arch," et cetera In a few examples the anterior limb has been obliterated in part or entirety, leaving a fibrous cord In most instances both of the limbs are patent and carry blood In very rare cases the two branches are equal in size In the majority, the anterior (left) arch is somewhat smaller than the posterior (right) one The division of the aortic arch into two channels implies that these limbs sur round some mediastinal structure In at least three instances the trachea alone has been encompassed, but in all others the esophagus as well as the trachea has been encircled

A double aortic arch may give rise to no symptoms, indeed, the majority of specimens which have been described have come from autopsy examination (or anatomical dissection) of elderly subjects who apparently had no important impairment of health from the malformation Such freedom from complications is not universal The presence or absence of symptoms will depend upon how much room is available between the two limbs of the aortic arch If sufficient space is present, it is obvious that the trachea will not be compressed, and the esophagus can displace forward during the act of deglutition However, if the "vascular ring"—if such it may be called—is small, there will be encroachment upon the esophagus and trachea If these complications appear, they are extremely apt to do so within the first year of life, and dysphagia or stridor become outstanding complaints Frequently, there is a recurring tracheo-bronchitis and indeed, death is prone to occur from superimposed pulmonary infection

Wolman[63] has very precisely described the clinical picture which is presented by these infants, and our observations are quite

similar to his Dysphagia and particularly stridor appear shortly after birth and are persistent These may be mild or they may be severe they can continue until death supervenes or surgical relief is instituted The respirations are noisy and wheezing and they often have a crowing quality Retraction in the suprasternal and intercostal spaces is usually found There may be a recurrent harsh or brassy nonproductive cough The cry is hoarse The respiratory rate may be markedly elevated When the child swallows fluid or solid food the respiratory noises are accentuated and at times mild or moderate cyanosis may appear Swallowing is slow and the baby might have to rest at frequent intervals to improve the breathing Infections of the tracheobronchial tree and the pulmonary parenchyma are presumably in some way stirred up or incited by the compression of the trachea they may also develop from spill over of food into the air passages

The roentgenologist[56] can recognize these abnormalities with a considerable degree of accuracy during life A swallow of barium permits visualization of the esophagus which may show little disturbance in the anterior posterior view but which will be displaced forward at the level of the third or fourth thoracic vertebra The size of the structure behind the esophagus may or may not be as large as that seen in a right aortic arch The filling defect on the posterior wall of the esophagus has a transverse direction and does not have the obliquity which will be described later in the section on anomalous right subclavian artery The trachea can be studied by antero posterior and lateral films of proper density It is usually possible to visualize the trachea because of its air content and to ascertain any deformity which is present If such visualization is unsatisfactory lipiodol can be inserted or sprayed into the larynx (without anesthesia) and excellent delineation of the trachea obtained By this means a lateral film will show compression of the anterior surface of the trachea just above the carina

Surgical therapy is now available for this abnormality even in the smallest of subjects It is impossible in most cases to divide the posterior aortic limb since this is almost always the larger of the two channels However by severing some portion of the anterior (left) limb the vascular ring can be broken and sufficient room can be made for the trachea and esophagus Just where the anterior limb should be divided will depend upon the size of the various portions of this limb having due consideration for maintaining an adequate flow to the vessels which might arise from this structure

The exact arrangement of the great vessels and the details of the operative procedure employed for alleviation of tracheal compression have been more fully presented in previous communications [16 52]

I have had very fortunate experiences with surgical treatment in two patients with these abnormalities The first was a nine months old child who first came to the hospital at four months of age with a complaint of wheezing respirations since birth Because of a widened superior mediastinal shadow which was thought

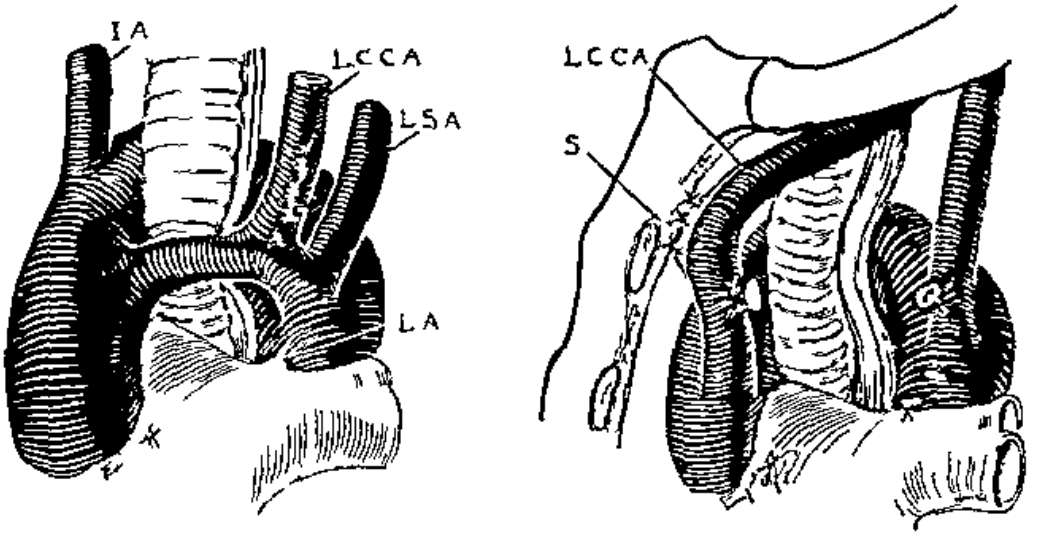


Fig 21 Sketch of double aortic arch from a five months old baby who had symptoms and roentgenographic evidence of compression of the esophagus and the trachea Double aortic arch as found at surgical exploration
Fig 22 Operative procedure carried out for double aortic arch shown
Fig 23 The anterior limb of the aorta has been divided between the origins of the left common carotid artery and the left subclavian artery The left common carotid artery has then been tacked to the back of the sternum so that it will not press on the anterior surface of the trachea By this procedure the child has been completely relieved of symptoms

to represent an enlarged thymus x ray irradiation was given without relief The child was hospitalized at three subsequent times for attacks of acute tracheobronchitis during each of which the baby was seriously ill The therapy on each occasion included the use of a steam room administration of oxygen appropriate chemotherapy and other supportive measures During these times the child was ill enough to be on the danger list With the care indicated, the baby survived each of these episodes of infection Finally suspecting some underlying anatomical abnormality the esophagus and trachea were visualized by roentgenographic means and changes were found as indicated in a preceding paragraph Exploration

was performed through a left antero lateral approach traversing the pleural cavity, and entering the mediastinum through its left side A double aortic arch was found, from the posterior limb of which came the innominate artery and from the anterior limb there arose the left common carotid and the left subclavian arteries It seemed best to divide the anterior aortic limb between the origins of the left common carotid and the left subclavian artery This immediately relieved the sterterous breathing and the respiratory sounds diminished almost to normal Unfortunately the left common carotid artery was left resting upon the trachea in a way which has given this child some residual symptoms Although greatly relieved from the preoperative state, the individual still has some persistent difficulties

The second patient was a five month old baby who entered the hospital because of stridor, cough, fever, and respiratory infection of two weeks duration The child was quite cyanotic and was in severe distress There was marked intercostal retraction He was placed in a steam room, given oxygen, penicillin and sulfadiazine For two days his condition was serious enough to require the use of various stimulants, but following this period he gradually improved and was discharged apparently free of infection on the nineteenth day Two months later there was again cough noisy, respirations, progressive anorexia, and weight loss Roentgenologic studies showed compression of the esophagus from behind and flattening of the trachea from the front At surgical exploration (Figure 21), again by way of the left pleural cavity, the superior mediastinum was dissected without difficulty and an abnormality was found which was precisely the same as that described in the last case The anterior aortic limb was divided as before, but in addition, the left common carotid artery was held forward and away from the trachea by anchoring it to the back of the sternum with several interrupted silk sutures (Figure 22) The respirations now became quiet and entirely normal The left lung was re expanded and the chest was closed Following operation the residual respiratory infection rapidly responded to treatment and the baby was discharged from the hospital on the twelfth day Since that time the child has been normal in every way He swallows without hesitation, has had no stridor or other respiratory symptoms of any kind There has been a rapid gain of weight and the parents are delighted with the result which has been obtained *

ANOMALOUS RIGHT SUBCLAVIAN ARTERY

"Dysphagia lusoria" is a condition in which hesitancy in swallowing occurs because of pressure on the esophagus by an anomalous right subclavian artery. In these patients the right subclavian artery, instead of arising in a normal way from the innominate artery, has an origin from the left side of the aortic arch so that the vessel must course upward and to the right crossing the midline to reach its normal exit on the right side of the thoracic cage. Holzapfel[6] made a study of 133 specimens and found that the artery ran behind the esophagus in 107 cases, between the esophagus and trachea in twenty, and in front of the trachea in six. The vessel usually crosses the midline of the body at about the level of the third dorsal vertebra. The first authentic report of this abnormality was more than two centuries ago, apparently by Hunauld in 1735. Particular attention was drawn to the malformation by an exceedingly well described and illustrated report of Bayford[64] in 1794. For many years he had attended a woman who had had marked difficulty in swallowing which had progressed to the point where she was emaciated and was in an advanced stage of starvation. She was removed to the county almshouse where she eventually died. At autopsy the esophagus itself was normal but it was indented by an anomalous right subclavian artery which arose from the left side of the aortic arch. To this clinical and pathological state he ascribed the name of *dysphagia lusoria*, indicating thereby that the dysphagia was due to a *lusus naturae (a freak or deception of nature)*.

An anomalous right subclavian artery does not necessarily give rise to symptoms; indeed it does not do so in the majority of instances. It may, however, press on the esophagus sufficiently so that the patient complains of hesitancy or discomfort during the act of deglutition. This may in no way impair the general health of the patient, and the symptoms might not increase during adult life. However, there are frequent observations to indicate that dysphagia lusoria may become more pronounced with advancing years. This aggravation of symptoms is related to increased rigidity of the great vessels in later life or to dilatation of the subclavian artery or the aortic arch in such a way that greater pressure is exerted on the esophageal tube.

The roentgenologist can now detect the presence of this abnormality with great certainty Visualization of the esophagus by a barium swallow shows that it is compressed usually on its posterior surface by some long narrow structure extending upward and to the right in an oblique direction The proportions of this filling defect correspond to that which one would suspect from the size of the subclavian artery The esophageal defect will be at the

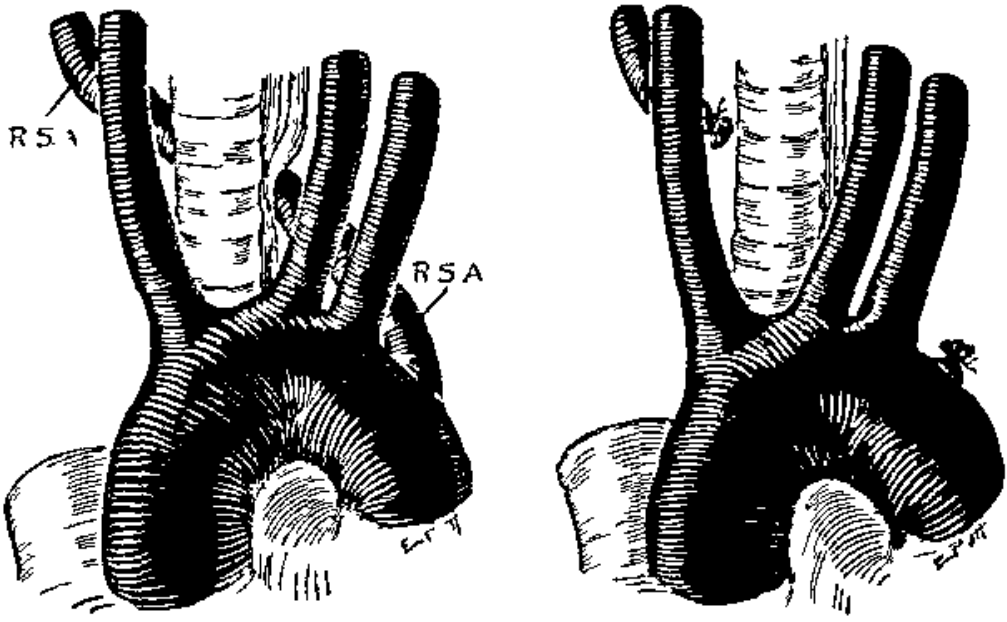


Fig 23 Sketch of an anomalous right subclavian artery which lacks its normal origin from the innominate artery but instead arises directly from the left side of the aortic arch Operative findings from a four months old baby who had difficulty in swallowing
Fig 24 Type of operation carried out for abnormality shown in Fig 23 The right subclavian artery was doubly ligated and divided so that it was completely displaced from its retroesophageal position

level of the third or fourth thoracic vertebra There is usually little or no dilatation of the esophagus above this area While the subclavian artery is usually on the posterior wall of the esophagus Neuhauser [6] has shown an example (unproved) of the vessel coursing between the esophagus and the trachea

It is possible to divide the first portion of a subclavian artery without serious impairment of blood flow to the corresponding arm Collateral channels which communicate with the second and third portions of the subclavian artery and with the axillary artery are sufficient to maintain an adequate flow of blood to the arm This has been extensively studied by Halsted and more recently has been substantiated by the series of cases reported by

Blalock[38] in which this vessel has been severed without any deleterious effects upon the arm These observations at once make it evident that a patient with dysphagia lusoria can be completely relieved of symptoms by division of the anomalous subclavian artery so that it is removed from the posterior mediastinum

We have had the opportunity to study the effects of such an operative procedure on a four month old infant who had had distress since about one month of age[66] Difficulty in swallowing increased since that time Whenever the child would attempt to suckle he would immediately stiffen up with pain and would cry After subsidence of this discomfort resumption of feeding would again bring on a spell of crying There was occasional regurgitation of milk At some feedings the child would take the full amount of formula but frequently he would not swallow more than an ounce or two In order to maintain the baby in a satisfactory state of nourishment it had become necessary to give him fifteen to twenty small feedings per day The general physical examination was negative Observation in the hospital showed that many feedings were taken well but at other times only a small amount of milk could be ingested After roentgenologic examination of the esophagus Dr Edward Neuhauser found a posterior esophageal defect such as above described The chest was explored through a left antero-lateral thoracic approach traversing the left pleural cavity (Figure 23) Without difficulty the superior mediastinum could be dissected and an anomalous right subclavian artery was found passing upward and to the right between the esophagus and the vertebral column The vessel was doubly ligated and divided in such a way that its distal end was allowed to retract to the patient s right beyond the esophagus (Figure 24) Following operation this child has had an extremely satisfactory course and has not had the slightest hesitancy in swallowing

While an anomalous right subclavian artery is a malformation which usually does not give rise to important symptoms there are some individuals who can obviously be helped by this surgical procedure which is not difficult to perform Whenever there is any serious impairment of health certainly this therapy should be advised Indeed when there are difficulties which do not necessarily endanger life but which are nevertheless quite disagreeable operation could bring comfort and relief

COARCTATION OF THE AORTA

Coarctation of the aorta is a narrowing or complete obstruction of the aorta. The lesion has been classified as a congenital one but the exact nature of the obliterative process is not entirely clear. The fact that coarctation appears in that part of the aorta adjacent to or near the ligamentum arteriosum suggests that the degenerative change which is concerned with closure of the ductus has in some way involved the aortic wall. There have been a few descriptions of coarctations which occurred in the abdominal or in the lower thoracic portions of the aorta. In most instances it is found in the distal part of the aortic arch or in the uppermost segment of the descending aorta so that it is near the ductus arteriosus or its obliterated remnant the ligamentum arteriosum. Coarctation has been said to occur about once in every 1000 to 1500 routine postmortem examinations. Pathologists have separated these abnormalities into two general groups in a way which is somewhat superficial but which has some practical importance. In the *infantile* type there is a very long segment throughout which the aorta is narrowed or obliterated. This narrowing is found in the aortic arch particularly in its distal one third or one half so that the left subclavian artery and possibly the left common carotid artery receive a poor supply of blood. This form is usually associated with other

Fig. 25. Specimen of coarctation of the aorta from a twenty five year old woman who died from intracranial hemorrhage. From Brainwell and Jones British Heart Journal 3 201 1941. The coarctation appears just beyond the origin of the left subclavian artery.

severe cardiac abnormalities and generally is incompatible with life for more than a few weeks or months In the *adult* type of coarctation the constriction is limited to a very short segment and appears just at or beyond the origin of the left subclavian artery (Figure 2) In this type the heart usually shows no congenital malformation or if such is present it is apt to be of a minor variety it is apt to be of a minor variety

Individuals with an adult type of coarctation have a variable prognosis regarding general health and longevity of life Occasional patients live to an advanced age and have little or no incapacitation but the majority of them develop complications of serious or even fatal significance (1) A localized or dissecting aneurysm may appear This may involve the aorta above the obstruction but frequently it is found in the lower aortic segment Such dilatation is *often on the basis of arteriosclerotic degeneration* but it can also appear in a wall which is free of arteriosclerosis and which is thin *because the media is deficient in elastic tissue* (2) *There may be* rupture of the upper or lower aortic segment and a sudden death *Rupture does not necessarily depend upon a pre-existing hyper* tension In a personal communication Dr Samuel Levine has told me of one woman twenty three years of age who was previously known to have a normal blood pressure in the arms but who had a sudden exodus from rupture of the aorta during the early part of pregnancy (3) The vascular abnormality may be the seat of superimposed infection especially with organisms of the *streptococcus viridans* type (4) The blood pressure in the upper part of the body may become elevated and with this may follow all of the ill effects and hazards of the hypertensive state Many of the fatalities in individuals with coarctation of the aorta are on the basis of cardiac failure or intracranial hemorrhage

In general coarctation of the aorta produces little or no disturbances in childhood and indeed it may go unnoticed Freedom from complications in this age period cannot be regarded as an indication that the individual will subsequently continue through life and be unmolested by the malformation which he possesses Indeed the interrogation of adult patients has usually impressed me with the fact that they look back upon their childhood or adolescent years as being free of symptoms and as having no limitation in physical activity or athletic endeavors As one reviews previous studies such as that made by Blackford[13] on 196 autopsied cases the serious nature of coarctation of the aorta is at once evident

More than 40 per cent of the individuals died between the ages of sixteen and thirty years This is in striking contrast to the low mortality rate in this age period—the prime of life—for the general population It is true of course that pathologic studies might tend to overemphasize the perils of coarctation because individuals who have died from it tend to be reported whereas those who have died from other causes might not be described in medical literature While it is freely admitted that some people may carry a coarctation into well advanced years this does not vitiate the fact that a high percentage of these patients are being cut off in early adult life because of the abnormality or one of its various complications

Coarctation of the aorta can be recognized with great ease The important point in its detection is the finding of an abnormal pressure relationship in the arms and legs Arterial pulsations in the femoral vessels (and in other arteries below this level) are greatly diminished or absent In normal subjects the systolic blood pressure of the legs should be twenty to forty millimeters of mercury higher than it is in the arms Whenever pressures of equal magnitude are found in the arms and legs, one can suspect a mild aortic block When the pressure in the legs is greatly below that in the arms one is certainly dealing with a high degree of aortic obstruction In many cases of coarctation the pressure in the legs is greatly reduced and no sounds can be heard when using the sphygmomanometer Hypertension may or may not exist in the arms In children and in a few fortunate adults the pressure in the arms may be within a normal range More commonly particularly in adults there is a moderate or marked elevation of diastolic and particularly of systolic pressure in the upper extremities The heart may be somewhat enlarged If collateral arterial channels have become well established many of these may be detected during careful physical examination Palpation over the antero-lateral portions of the chest below the breast area may indicate pulsating intercostal arteries Sometimes palpation in the axilla will show pulsation of vessels along the course of the long thoracic nerve Most frequently pulsations can be felt and sometimes seen just below and medial to the lower tip of either scapula These represent greatly enlarged arteries within the substance of the trapezius or the latissimus dorsi muscles The arteries at the base of the neck may have a heaving pulsation Murmurs are somewhat variable most commonly there is a systolic murmur of moderate intensity heard best over the left upper portion of the precordium but it is

fairly well transmitted to the back particularly to the left of the spine If the aortic blockage is complete there may be no murmur at all Auscultation may reveal systolic murmurs of other anomalies such as an interventricular septal defect or there may be a continuous murmur in the pulmonic region suggestive of a patent ductus arteriosus Furthermore large and tortuous collateral arteries can

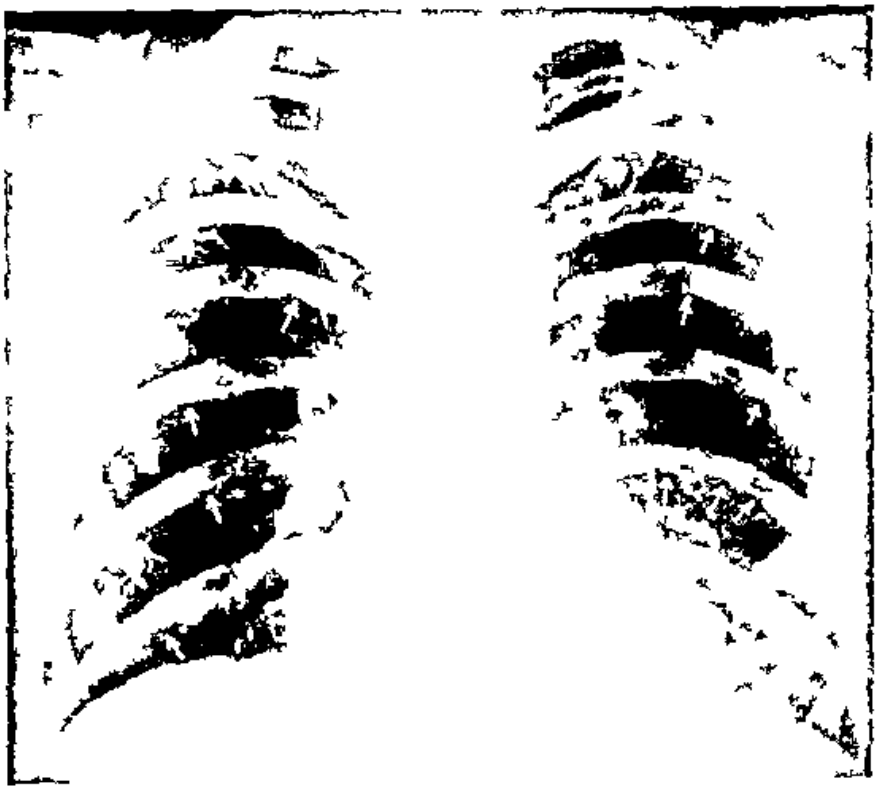


Fig 26 Roentgenogram of a thirty year old man with a proved coarctation of the aorta The heart is moderately enlarged The aortic knob is smaller than normal There is notching of the inferior borders of the ribs some of which are indicated by arrows

give rise to systolic or even continuous murmurs particularly over the scapular regions of the back

Roentgenologic examination (Figure 26) may show some hypertrophy of the heart particularly if there is associated hypertension The aortic knob may be smaller than normal or there may be a lack of fullness in the aorta in that portion which represents the junction between the aortic arch and the descending aorta Of diagnostic importance is the notching of the inferior borders of the ribs These occur in the posterior or postero-lateral parts of the ribs and represent erosions of bone by tortuous and pulsating subjacent intercostal arteries They usually do not appear in the upper two or three ribs or in the lower few ribs It is exceedingly

rule for them to be found before seven or eight years of age but Neuhauser[55] has described them in a baby nine months old It is possible to visualize the aortic arch and the aorta down to the point of obstruction by the intravenous injection of 70 per cent diodrast and taking a chest film at the appropriate time after the radio opaque material has circulated through the lungs and heart and has been delivered into the first portion of the arterial system We have clearly identified coaretation of the aorta by this means

Individuals with coaretation present variable complaints In babies there may be no symptoms and the condition may be de tected only by routine examination In some instances the murmur may draw the physician s attention to the existing defect In the childhood years there may be epistaxis or mild headaches In the teen ages headaches may become more prominent, and the subject may be conscious of a heart beat which is unduly forceful One man of twenty volunteered the information that his feet and lower legs were frequently cold but that simultaneously his head and shoulders would be hot and flushed He likewise noted that moder ate exercise such as running upstairs would produce weakness in his legs but that there was concurrent pounding in his head and this was frequently followed by an epistaxis As a rule these indi viduals are very well developed in their physical stature indeed many of them appear to be above average in development par ticularly in the upper part of the body Patients beyond twenty or twenty five years of age commonly notice a diminished tolerance for exercise Not infrequently patients in the late twenties and beyond have symptoms of frank cardiac failure Pain in the back should make one strongly suspect the presence of an aneurysm or at least of dilatation of one of the aortic segments and possibly impending rupture Pregnancy may be tolerated fairly well yet the gravid state and the increased demands imposed by the placental circulation appear to be a great hazard for the individual with an obstruction in the main arterial pathway Mentioned above is one woman of twenty three who died of aortic rupture during early pregnancy I have had occasion to examine a second woman who went through an initial pregnancy fairly well but during a second one at the age of twenty four, she developed left sided cardiac failure, from which she presumably will not recover

An intensive study of the problem leaves little doubt of the hazards which must be faced by an individual with coaretation of the aorta It is evident that this is a cardiovascular abnormality

of a serious sort which should excite attempts to bring relief by surgical means Four general operative approaches seem possible (1) For those individuals with hypertension, an extensive sympathectomy of the Smithwick type is said to diminish the pressure, but we are yet not certain how long these beneficial effects will last (2) The aortic obstruction could be by passed by severing the left subclavian artery at the base of the neck, and turning its proximal end downward and anastomosing it to the aorta below the obstruction This general principle has been suggested by Blalock and Park [79] and its feasibility has been demonstrated by work upon dogs The efficiency of such a procedure in man is somewhat doubtful because the anastomatic channel thus established would probably be of insufficient size Furthermore, the disruption of the subclavian artery would cut off many important collateral channels which invariably come from this vessel and its branches (3) The obstruction might be removed from the aorta and a segment of vein or aorta from another subject implanted therein by direct suture or by use of the Blakemore [74] technique (4) The constricted part of the aorta might be removed and the aortic continuity re established by anastomsis of the free ends of the vessel It is along this line of attack that all of my efforts have been directed The feasibility of such a procedure was demonstrated by the animal experimentation of Gross and Hufnagel [82] It was performed by Crafoord [80] and has been further established by our experiences in seven cases as noted below

Experimental Observations

From the laboratory we wanted to gather information regarding two general questions First, could the upper part of the descending aorta be cut in half and its ends re anastomosed with any reasonable degree of security? Second could a portion of aorta one or two centimeters in length, be removed from the upper end of the descending aorta, and was there sufficient elasticity in the remaining parts of the vessel to allow the ends to come together for establishment of a direct anastomosis? These experiments were begun in 1938 and met with numerous adversities Because of other duties they were suspended during the war I was later joined by Dr Charles Hufnagel, who was exceedingly helpful in the work, and for whose assistance I am very grateful A number of animals, which had been sacrificed for other reasons, were used to practice steps and to familiarize ourselves with the technical problems which

TABLE I. DATA ON DOGS UNDERGOING DIVISION AND SUTURE OF THE AORTA

Dog No	Date of Operation	Date of Death	Cause of Death	Condition of Suture Line	Segment of Aorta Excised
1	3-13-38	3-14-38	Never regained consciousness (possible anesthetic death)*	Satisfactory	None
2	3-22 39	3-22-39	Cardiac failure on removal of aortic clamps	Satisfactory	None
3	3-21-44	3-22-44	Never regained consciousness (possible anesthetic death)*	Satisfactory	None
4	4-17-44	4-17-44	Anesthesia	Satisfactory	None
5	4- 2-44	5- 7-44	Empyema	Satisfactory	None
6	5- 9-44	5-10-44	Hemorrhage	Hemorrhage from suture line (clot in lumen)	1 0 cm
7	5-19-44	1-11 45	Distemper	Satisfactory	None
8	5-26-44	5-26-44	Never regained consciousness (possible anesthetic death)*	Hemorrhage from suture line (clot in lumen)	None
9	6- 9-44	6- 9-44	Never regained consciousness (possible anesthetic death)*	Satisfactory (clot filled the lumen)	None
10	6-14 44	6-15-45	Sacrificed	Satisfactory	2 0 cm
11	6-16-44	6-19-44	Hemorrhage from chest wall	Satisfactory	None
12	6-22-44	6-26-44	Delayed hemorrhage from suture line	Late hemorrhage from suture line	None
13	1-15-45	1-27-45	Pneumonia	Satisfactory	None
14	3-16-45	6-15-45	Sacrificed	Satisfactory	1 5 cm
15	4-18-45	4-24-45	Unknown	Satisfactory	2 5 cm
16	5-31-45	5-31-45	Sacrificed (at completion of operation)	Satisfactory	None
17	5-31-45	5-31-45	Sacrificed (at completion of operation)	Satisfactory	None

*On autopsy, no adequate cause for death could be found in these animals It is possible that intracranial disturbances during the period of aortic occlusion contributed to these fatalities

might be encountered in the living dog In very young or small animals, the aorta was so thin that it did not appear to have sufficient substance or toughness to permit manipulating it with any degree of safety In larger animals, beyond 25 or 30 pounds in weight, the aorta was usually thick enough to be handled with ease Various types of anastomoses were tried, most of them need not be considered here because they were thought to be unreliable A type of union which seemed to be far superior to all others is that which is indicated in Figure 27 The stitches are placed in such a way that the thread passes through entire thickness of aortic wall It is a continuous mattress suture which turns the ends of the aorta outward and brings intima to intima

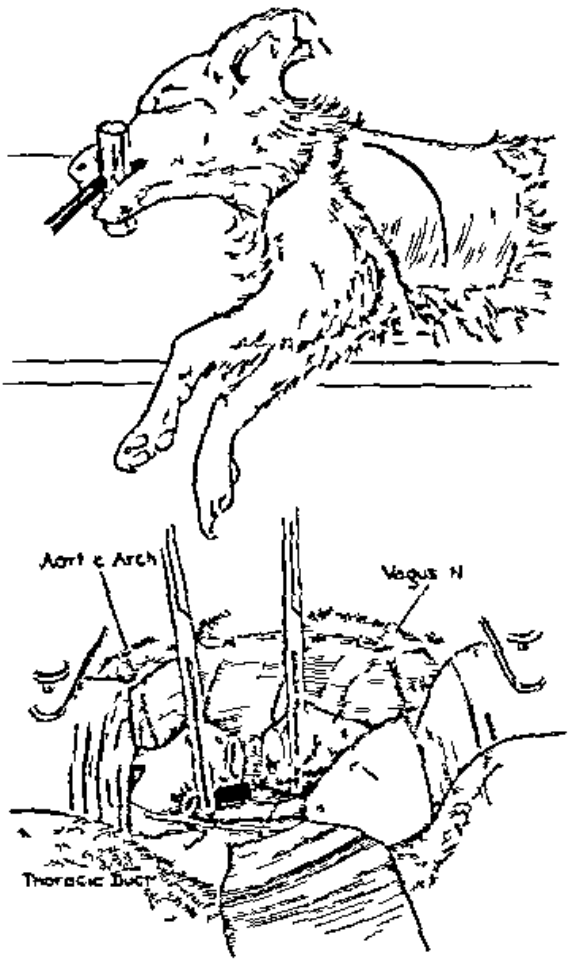


Fig 27 (A) Experimental procedures employed for practicing operation on the upper thoracic aorta of dogs *Above*— Exposure through a left postero lateral incision. *Below* — Chest opened, with transpleural approach to the aorta *Clamps have been applied to the vessel* and the aorta has been divided.

Much to our surprise and delight, it was found that a short segment of aorta could be removed and that an aortic reconstruction could still be performed When a segment was thus excised the ends of the aorta retracted from one another in a most distressing way However, the remaining vessel was elastic enough so that approximation of the holding clamps would bring the vessel ends together and permit a direct suture without tension while the anastomosis was being done

After practicing these steps on dog cadavers, living animals were employed and our convictions were confirmed that the aorta could be divided, that a short segment of it could be removed, and

that the continuity of the vessel could be re established (Figure 27) Some data from seventeen animals are included in the accompanying table

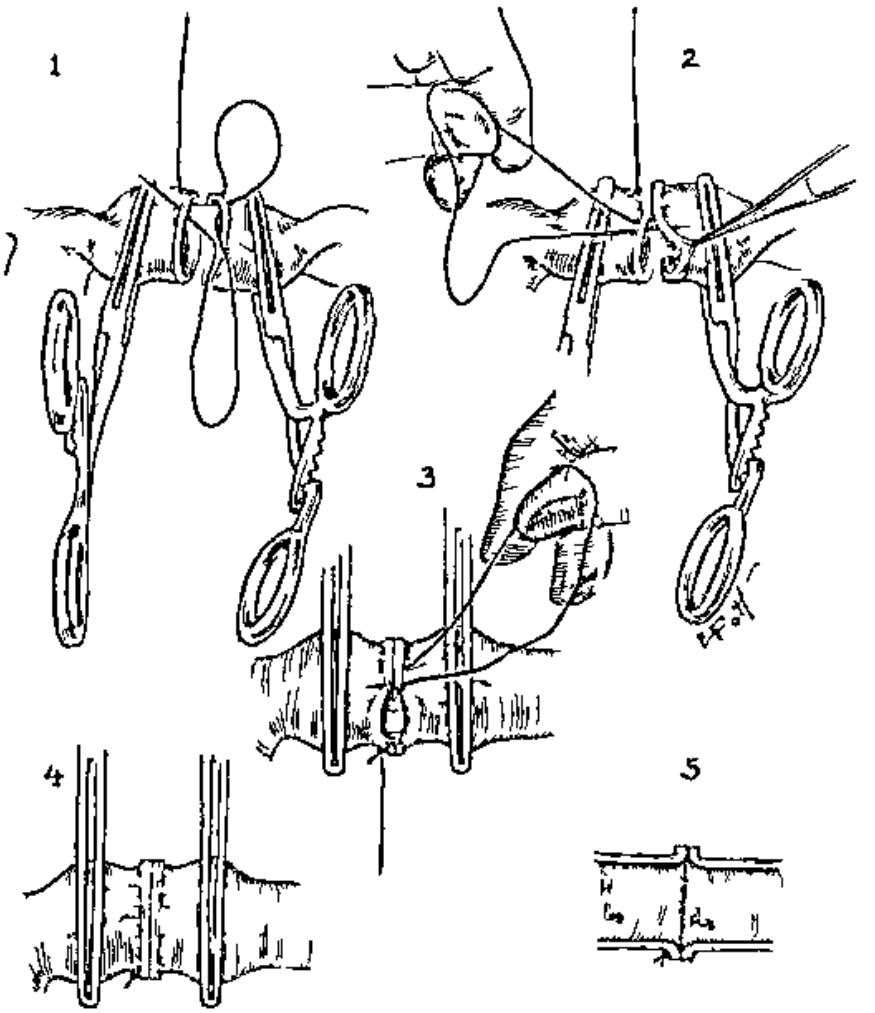


Fig 2 (B) Method of reconstruction of aorta The clamps are held by first assistant so that the aortic ends are brought together and there is no tension on the suture line during establishment of the anastomosis (1) Aorta rotated forward and suture begun on the back wall (2) Suture continued Each stitch passes through the entire thickness of the aortic wall as a continuous mattress everting stitch (3) Back wall of aorta sutured and the aorta has now been untwisted into its normal alignment Stitch is being continued anteriorly (4) Suture completed Ends of the aorta are everted (5) Cross sectional view of method of bringing intima to intima and turning the ends of the aorta outward

Some remarks might be made which are pertinent to our laboratory studies Of prime importance in reconstructing an aortic tube so that there would be no subsequent hemorrhage was the meticulous apposition of the vessel ends in such a way that every stitch was

placed with extreme care, and that each individual stitch was drawn up with just the right tension If the anastomosis was properly performed, there was little or no leakage from the suture line when the clamps were removed and there was very little danger of hemorrhage subsequent to the operation Furthermore, it was evi dent that the local use of hemostatic agents or packs around the suture line was a very poor and unreliable substitute for an accur

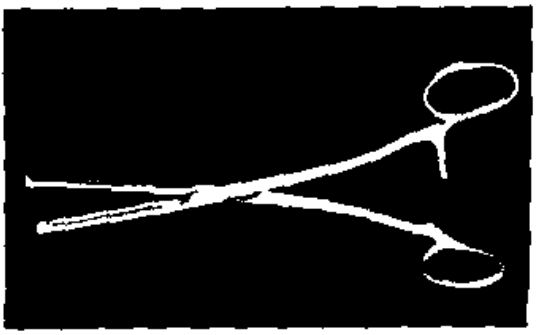


Fig 28 Instrument improvised for clamping the aorta The jaws are stout enough to give complete hemostasis and yet they have sufficient resilience so that they do not crush the aortic wall The instrument is used without rubbers

Fig 29 Specimen from aortic arch and thoracic aorta of a dog which *was sacrificed three months after* ex cision of 1 5 cm of the aorta and en l to en l suture of the vessel Arrows show the operative site The healing *has been excellent*

ate anastomosis Three dogs died of hemorrhage, one on the day of operation, one on the follow ing day and the last on the fourth postoperative day It was our impression that such bleed ing was due to faulty techniques and that with increasing experi ence this complication could be largely avoided

In none of these dogs was heparin or dicumarol employed fol lowing operation In three animals there was some clot within the lumen at the site of the anastomosis *In three others there were* one or more pin head sized thrombi on the suture line In general it was felt that clotting at the site of anastomosis was not an im portant obstacle to the performance of these anastomoses

The total obstruction of the aorta, as was necessitated by the application of clamps, naturally raised certain questions concern ing the ability of the heart to withstand such a measure In general,

clamping of the aorta below the left subclavian artery had little effect upon the cardiac mechanism In some animals the heart would speed up slightly or would dilate to a minor degree but in no instance did the heart stop or give evidence of any important embarrassment After performance of an anastomosis removal of the clamps was apt to produce serious changes in the cardiac rate or activity and in one animal there was immediate cardiac failure However when the clamps were removed slowly from the aorta circulatory adjustments could be made in a more gradual manner and were completely satisfactory

Double clamps were placed upon the aorta above and below the region which was to be transected or excised, these instruments having the two fold function of affording hemostasis and also of providing handles by which the ends of the aorta could be pushed toward one another while the steps of suture were being performed A number of standard instruments were tested to study the requirements imposed by this operation Any rubber covered instrument was wholly unsuitable because of the possibility of slippage on the aortic wall Kelly clamps full length clamps Kocher clamps et cetera had too much of a crushing effect upon the vessel wall and appeared to be dangerous Furthermore they often grasped the side of the vessel which was toward the base of the jaws but simultaneously might not grasp adequately the opposite side of the wall which was toward the tip of the clamp Bethune tourniquets were fairly useful but they wrinkled the stump of aortic wall so that it was often difficult to sew the ends of the vessel Finally two instruments (Figure 28) were revamped from Moynihan enterostomy clamps and these gave great satisfaction The ends were sawed off and the ends fitted with an interlocking peg so that the jaws would not wiggle sideways when closed The longitudinal slit in the jaws insured against any side-slipping of the clamps on the grasped aorta Cross markings were filed on the jaws to preclude end slipping The jaws were just springy enough so that they would clutch an aorta without crushing it Post mortem studies have not shown any important damage to the aortic intima or to the outer wall at the sites where these clamps had been used

A serious complication of these operations appeared in the form of hind leg paralysis in some dogs Spinal cord examinations showed diffuse degenerative changes which had apparently resulted from local ischemia We were reasonably certain that such ischemia was

not present following operation, because these animals had good femoral pulsations during the postoperative period, and there was an adequate lumen to the aortas when examined post mortem Hence, we could not escape the conclusion that the spinal cord damage occurred while the aorta was temporarily obstructed by clamps during the operation To study further the effects of temporary obstruction of the aorta, especially in relation to spinal cord degeneration and hind leg paralysis, twenty additional dogs were operated upon by opening the chest, placing a clamp on the upper thoracic aorta for a given period, and then releasing the clamp The dogs were kept and observed for periods varying from one week to two months following operation These animals, combined with the seventeen which had been operated upon primarily for severance and suture of the aorta, provided a group of thirty seven in which the aorta had been obstructed for periods varying from four minutes up to one hour each Some dogs with occlusion of the aorta for forty five minutes or fifty minutes developed no hind leg paralysis, whereas others with shorter obstruction showed definite neurological damage In no instance did a paralysis develop when the aorta had been obstructed for less than ten minutes Colson,[75] Carrel,[6] and Blalock and Park[75] have previously commented upon the correlation of temporary aortic obstruction and hind leg paralysis Their remarks, and our personal observations, at first made us fear that neurological changes in the spinal cord would prohibit any operations on the aorta of man—procedures which could not possibly be completed in less than ten minutes However, it is highly important to point out that when aortic operations are performed in human subjects, there is extremely little likelihood of neurologic complications from temporary aortic obstruction, because adequate collateral channels have been established prior to operation

Operations in Man

To date, I have operated upon seven humans for coarctation of the aorta Some notes on these experiences, particularly the technical aspects of operation, have been recorded elsewhere[53 54] In all of these cases exposure has been through the back (Figure 30), using a long, curvilinear incision running from the level of the second thoracic vertebra downward and outward along the medial border of the scapula toward the posterior axillary line Traversing

the great muscles is apt to be very slow and tedious because within the substance of the trapezius the latissimus dorsi and the rhomboid muscles there are large collateral arterial channels which make the field exceedingly vascular and which require a great deal of clamping and ligation Eventually the scapula can be freed from the chest wall and pulled upward and outward to expose the costal cage The posterior half of the fifth rib is removed subperiosteally Short segments can also be taken from the angles of the fourth sixth and seventh ribs The operator is ever conscious of ooze from the intercostal muscles or from various other structures which indicate the vascularity of the chest wall The chest is entered throughout the bed of the fifth rib and the wound is then extended upward and downward near the vertebral column by cutting through the adjacent intercostal muscle bundles With suitable self retaining retractors a fairly good exposure can be obtained The desired portion of the aorta is now brought into view by splitting open its encasing parietal pleura In the fifth case of this series the coarctation was just opposite the origin of the left subclavian artery but in all other instances the constricted zone was one or two centimeters below this vessel and immediately opposite the ligamentum arteriosum (or a patent ductus arteriosus in one case) The external constriction of the vessel may be quite apparent and deep In three instances the outer surface was only slightly indented but palpation of the vessel wall in this region revealed an excessive thickening so that the lumen of the aorta was obviously no more than two or three milli

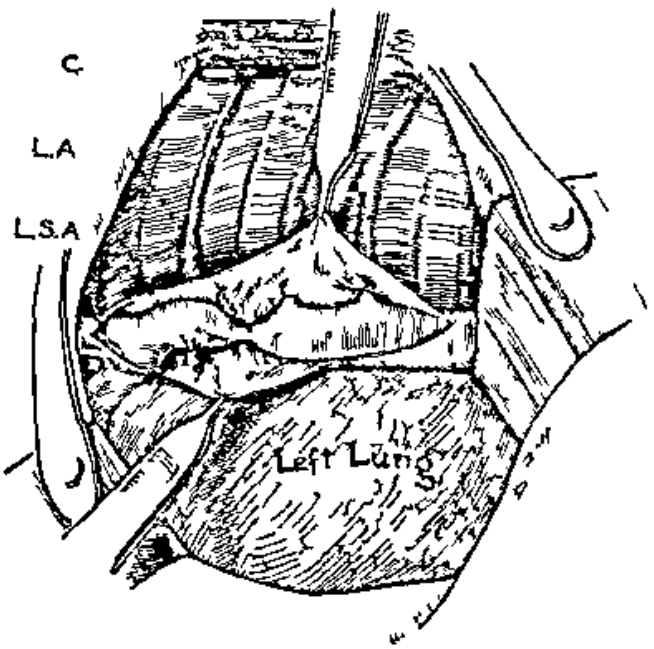


Fig 30 (A) Sketch made at operation on a patient with coarctation of the aorta Left postero lateral approach through the pleural cavity Coarctation at C L A Ligamentum arteriosum. LSA left subclavian artery The intercostal vessels are greatly enlarged The aorta above the constriction had a heaving pulsation whereas the vessel below this stenosed did not have an intrinsic pulsation.

meters in diameter The aorta above the constriction and the great vessels which arise from the arch show a heavy pulsation which is more marked than normal The aorta below the constriction has little or no intrinsic pulsation The intercostal arteries are extremely large, and may be five to seven millimeters in diameter They are usually tortuous in many places and are apt to have thin walls

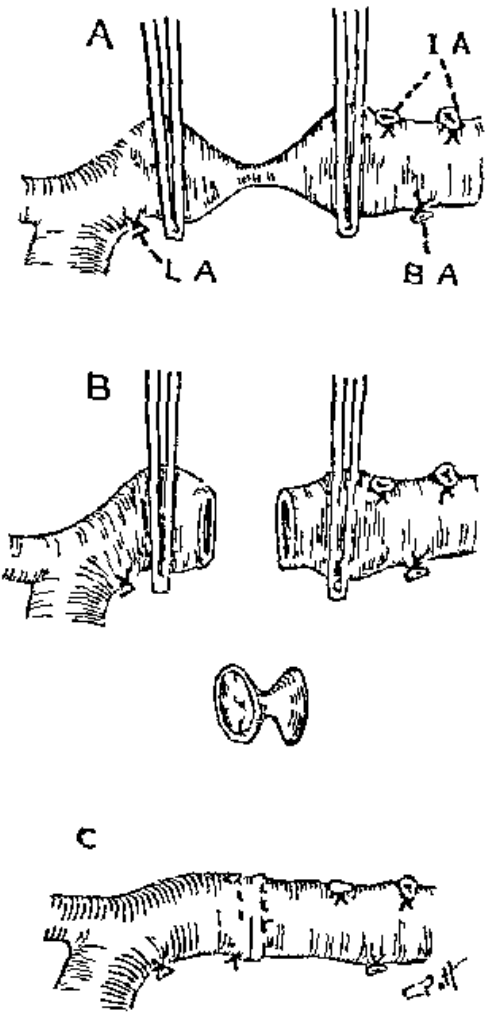


Fig 30 (B) A Segment of aorta 5 to 6 cm in length raised from its bed Intercostal arteries IA, have been doubly ligated and divided Bronchial artery, BA, has been doubly ligated and severed The ligamentum arteriosum, LA, has been cut B Clamps applied to aorta and narrowed segment has been excised C Reconstruction of the aorta by end to-end anastomosis, by technique shown in Fig 27

In order to free up a segment of aorta five to six centimeters in length it is necessary to doubly ligate and divide at least two sets of intercostal arteries below the constriction, to divide bronchial arteries when they exist, and to divide the ligamentum arteriosum (or the ductus arteriosus) This dissection must be extremely slow and careful for fear of injuring one of the vessels and setting up uncontrollable bleeding Care should be exercised about putting undue strain on an intercostal artery at its junction with the aorta since this is a point of considerable anatomical weakness When a portion of the aorta has been dissected from its bed a linen tape can be passed around it to facilitate the subsequent handling of it, and to raise it up for further severance of any adhesions or small vessels which might lie on its under surface Dissection posterior to the aorta must be carried very close to the vessel, to leave the thoracic duct undisturbed The vagus and recurrent laryngeal nerves can be displaced forward so that they are out of harm's way

From here on, the steps in treatment of coarctation of the aorta

are precisely the same as those which were practiced on dogs for excision of a segment of the vessel and re anastomosis of its remaining ends With the clamps in place a segment one to one and

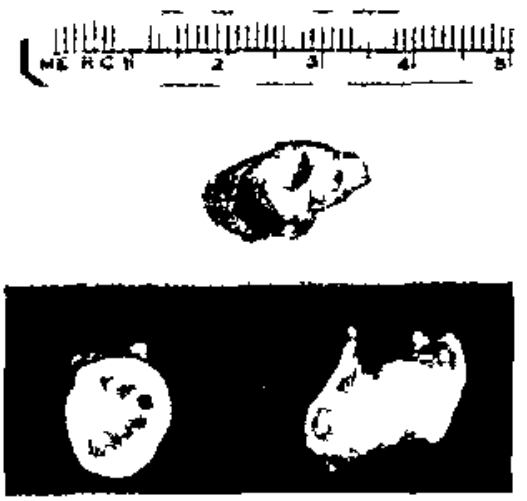


Fig 31 Two specimens removed from patients with coarctation of the aorta In each specimen the lumen was only 2 3 mm in diameter

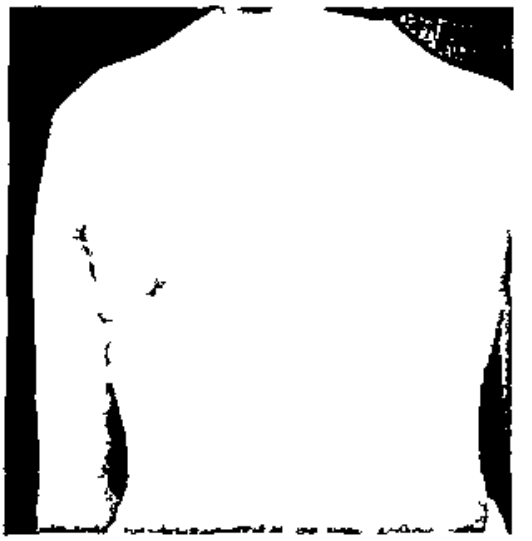


Fig 32 Post operative photograph showing position of wound on back following surgical treatment of coarctation of aorta

a half centimeters in length is cut away It is highly important to have a first assistant who has practiced the necessary steps so that he can manipulate the aortic clamps, and bring the ends of the aorta together so that they can be sutured without the slightest tension

In the first patient who was operated upon a boy of six years a very satisfactory anastomosis was established but on removal of the clamps the child promptly died I assume that this was a form of shock brought about by the sudden release of blood into the lower part of the body, where it pooled and could not return with sufficient rapidity to supply the heart with a circulating medium In the six subsequent operations three precautions were taken to avoid such a catastrophe (1) The last clamp was removed from the aorta very slowly (over a period of five to ten minutes) (2) The patient was tipped into a moderate Trendelenburg position (3) Several hundred cubic centimeters of blood were infused into an ankle vein By these measures the heart was supplied with an adequate amount of blood and the readjustments in the circulation were made without anxiety

The first five patients were individuals from six to sixteen years

of age and in each instance the operative procedure though long and difficult was carried out satisfactorily and with a feeling that an aortic tube had been reconstructed that was strong which was adequate in size and which had every promise of functioning in a normal way In the sixth patient a man of thirty who weighed nearly 200 pounds the exposure through the back was not all that could be desired and the anastomosis had to be performed deep *in a hole where it was exceedingly difficult to place the stitches* accurately Furthermore the lower aortic segment was extremely thin and about three times the diameter of the upper portion of the aorta Openings of a similar size could be fashioned in the two segments but while performing the anastomosis it was obvious that the lower vessel was thin and friable and that the stitches would cut through it frequently and had little holding power I had no feeling of security when the anastomosis was finally completed The patient was in a satisfactory condition for twenty four hours after operation but he then suddenly expired presumably from hemorrhage at the suture line No autopsy was performed

The seventh case a male of twenty years weighing 190 pounds who had an exceedingly muscular development again presented difficulties at the operating table which were almost insurmountable The exposure through the chest wall was again somewhat limited Most disturbing was the fact that good mobility could be obtained for the lower thoracic segment but the upper portion—the aortic arch—was completely unyielding and could not be raised from its bed as it had been in all of the previous patients I assume that the fixation of the arch was related to the long standing high degree of hypertension The limited mobility of the upper segment prevented turning it adequately to bring it into optimum position for performance of an anastomosis The line of suture was quite unsatisfactory and it was necessary to re enforce it with a number of extra stitches grasping the adventitia and media in a way which unfortunately inverted and constricted the vessel at the site of the anastomosis Thus produced a lumen which was but eight or nine millimeters in diameter and which was subsequently found to be insufficient for relieving the hypertension

The disappointments in these last two cases make me feel that these operations for correction of the aorta should probably be employed only for younger individuals—possibly up to sixteen years of age However it is possible that an adult who has a thin chest and poorly developed musculature will present more favor

able circumstances for operation than was the case in my last two patients In young subjects the procedure can probably be completed with a risk which is not too high but in older individuals the surgical risks are probably enormous and should not be under

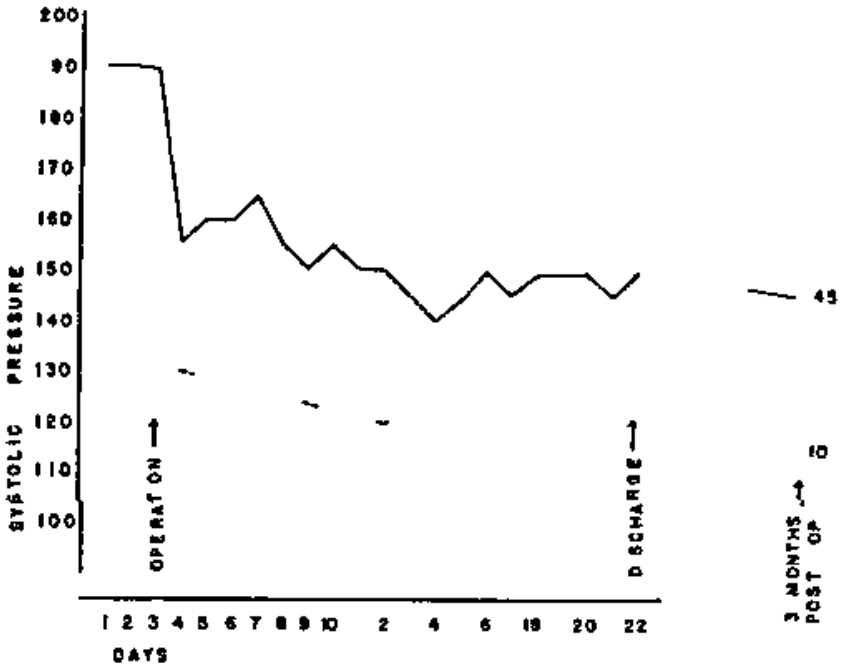


Fig 23 Blood pressure chart from patient 2 Solid line indicates pressures in arms Dotted line pressures in legs Prior to operation no pulsations could be felt in the legs During the ten days following operation there has been a decline in the arm pressures

taken until we have had further chance to gain more operative experience in the treatment of this condition

Comments Upon Successfully Treated Cases

Our series includes seven patients operated upon for coaretation of the aorta The ages of these were respectively six twelve ten sixteen, eleven thirty and twenty years The histories of these patients included a wide variety of symptoms Most of them had epistaxis at one time or another Most of them had headaches though this was not an outstanding symptom Several had palpitation Three complained of coldness of the legs Three noticed some weakness of the legs after moderate exercise One child was quite irritable and difficult to control The sixth patient complained primarily of pain in the back of several months duration The seventh individual had noticed a marked diminution of tolerance to exercise There was no question about the diagnosis in any of

these patients since there was a marked disparity in pressures of the arms and legs as indicated in the accompanying table

TABLE II DATA FROM PATIENTS OPERATED ON FOR COARCTATION

Case	Age	Sex	Representative Pre-Operative Systolic Pressure in Arms	Pre-Operative Systolic Pressure in Legs
1	6	M	170 mm of Hg	Unobtainable
2	12	F	215 mm of Hg	Unobtainable
3	10	M	155 mm of Hg	Unobtainable
4	16	M	175 mm of Hg	Unobtainable
5	11	F	155 mm of Hg	Unobtainable
6	30	M	220 mm of Hg	? 110
7	20	M	215 mm of Hg	Unobtainable

In the first of these patients there was a fatality on the operating table, after a very satisfactory aortic anastomosis had been obtained The clamps were removed too quickly from the aorta

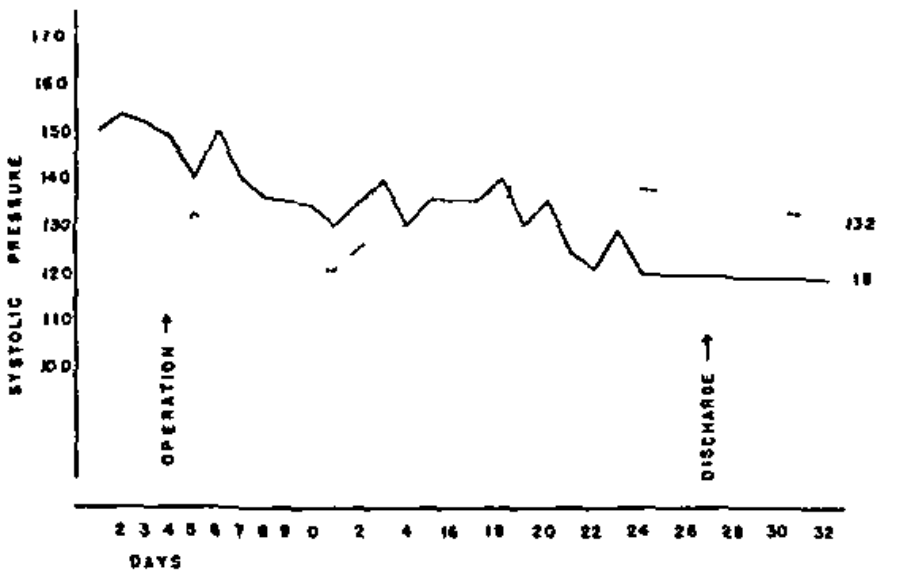


Fig 34 Blood pressure chart from patient 3 Solid line pressure in arms, dotted line, pressure in legs

This error has been corrected in all subsequent cases and this should not prove to be any source of trouble in future operations The sixth and seventh patients were both very large men, who presented formidable obstacles from the point of view of gaining adequate exposure, a problem which is not insuperable In addition, both of

them had some peculiarity in the aorta itself—one with a thinning and dilatation of the lower aortic segment and the other with a striking fixation of the aortic arch—which are probably situations which cannot be overcome by increased operative experience Just

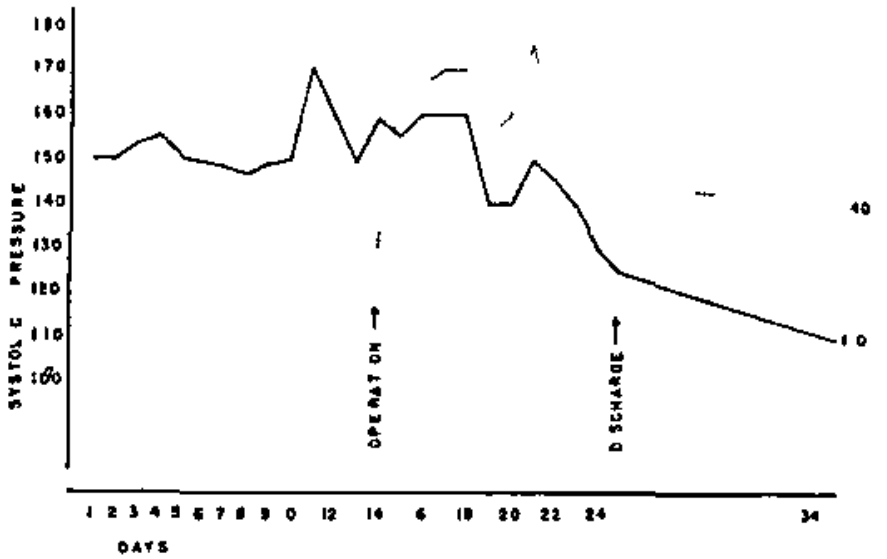


Fig 35 Blood pressure chart from patient 4 Solid line, pressure in arms Dotted line, pressure in legs Concurrent with the post operative appearance of pulsations and demonstrable pressures in the legs, there has been a post operative fall in the pressures in the arms

how frequently these disturbing factors will be found in adult patients, we are as yet unable to say While they have quelled my enthusiasm for further attempts at this operation in adults, it should be pointed out that Crafoord has operated upon one man of twenty seven with very satisfactory results However, my disappointing experiences serve to support a personal opinion that operations of this sort will primarily be of benefit to individuals who are young, whose symptoms are still mild, and who have not yet developed serious complications

In patient 7, there has been only a very slight fall in the blood pressure following operation, a fact which is obviously related to the establishment of too small a lumen at the site of the anastomosis In patients 2, 3, 4, and 5, there have been striking changes in the blood pressures as indicated in Figures 33, 34, 35, and 36 In none of these four patients was any pulsation felt in the femoral arteries, dorsalis pedis arteries, or the popliteal arteries before operation, nor could any sounds be heard when the sphygmomanometer was attached to the leg Following operation all four of these individuals have a very satisfactory pulsation in the

femoral arteries and other vessels of the legs They all have readily demonstrable blood pressure readings as shown by the dotted lines on the charts Of some interest is the fact that the blood pressure in the arms did not fall precepitously at operation or in a few hours

Fig 16 Blood pressure chart from patient 5 Solid line systolic pressures in *arms Dotted line systolic pressures in legs Prior to operation no sound could* be heard in the legs with the sphygmomanometer The reduction in pressures in the arms occurred gradually over a period of about two weeks

thereafter The diminution in arterial pressure of the arms took place over a period of ten to twelve days Hence it was assumed *that the vascular bed in the lower part of the body was small* because it had not been subjected to normal pressures prior to operation It is also possible that the peripheral bed had an increased vascular tone, dependent upon an over activity of the sympathetic apparatus *I do not know which of the two explanations is the* proper one but suffice it to say that apparently a period of time is required to dilate the vascular bed and that simultaneous with this change, the pressure in the upper extremities will fall

The extent of our observations is limited yet the findings clearly indicate that increased pressures in the upper part of the body can be greatly reduced by a surgical procedure which removes the obstructed segment of aorta *I would like to add that these operations* should not be undertaken lightly, and should be contemplated only by those who have had considerable experience in thoracic surgery, who are well acquainted with the techniques of blood vessel suture, and who are *willing to practice the necessary steps*

in the experimental laboratory These operations are lengthy and are exhausting for the surgeon and his team yet they are exceedingly gratifying because they bring a bright ray of hope to young individuals who have a serious vascular disorder which has heretofore carried a rather grave prognosis

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THIS BOOK

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by

ROBERT E GROSS M D

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